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A Neuropsychological Investigation with Adult Downs Syndrome Subjects

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A NEUROPSYCHOLOGICAL INVESTIGATION WITH
ADULT DOWNS SYNDROME SUBJECTS

by
Nancy Beard Scholle

A Dissertation Submitted to the Faculty of the
Graduate School of Loyola University of Chicago
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VITA

The author, Nancy Beard Scholle, was born on August 9, 1948, in Westminster, Maryland.

She received her elementary education in the public and parochial schools of Westminster, Maryland, and her secondary education at Villa Regina Academy in Baltimore, Maryland, where she graduated in 1966. She received her degree of Bachelor of Arts in August, 1970 from the College of Notre Dame of Maryland, with a major in Social Welfare. In June, 1973 she was granted her Master of Arts degree in Psychology from Roosevelt University in Chicago, Illinois. In 1978 she began studies for her Doctorate in Clinical Psychology at Loyola University in Chicago, Illinois. She completed training at the Loyola Child Guidance Center and at the Veterans Administration Hospital in North Chicago, Illinois.

She has worked professionally as a school social worker and school psychologist in facilities for mentally handicapped and emotionally disturbed children and adults. In 1974 she received certification as an Illinois School Psychologist. She has also presented professional papers at the Council for Exceptional Children Convention and the American Association on Mental Deficiency Convention.
TABLE OF CONTENTS

<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>ACKNOWLEDGMENTS</td>
<td>ii</td>
</tr>
<tr>
<td>VITA</td>
<td>iii</td>
</tr>
<tr>
<td>LIST OF TABLES</td>
<td>vi</td>
</tr>
<tr>
<td>LIST OF FIGURES</td>
<td>vii</td>
</tr>
<tr>
<td>INTRODUCTION</td>
<td>1</td>
</tr>
<tr>
<td>REVIEW OF RELATED LITERATURE</td>
<td>5</td>
</tr>
<tr>
<td>The Nature of Downs Syndrome</td>
<td>5</td>
</tr>
<tr>
<td>Discovery and Incidence</td>
<td>5</td>
</tr>
<tr>
<td>Intellectual Subnormality</td>
<td>7</td>
</tr>
<tr>
<td>Language Disorders</td>
<td>9</td>
</tr>
<tr>
<td>Physical and Neurological Abnormalities</td>
<td>16</td>
</tr>
<tr>
<td>Personality Abnormalities</td>
<td>25</td>
</tr>
<tr>
<td>Summary</td>
<td>27</td>
</tr>
<tr>
<td>Human Neuropsychological Testing</td>
<td>28</td>
</tr>
<tr>
<td>History of Human Neuropsychological Theory</td>
<td>28</td>
</tr>
<tr>
<td>The Halstead-Reitan Approach</td>
<td>32</td>
</tr>
<tr>
<td>The Luria Approach</td>
<td>34</td>
</tr>
<tr>
<td>Standardization of the Luria Approach</td>
<td>40</td>
</tr>
<tr>
<td>Neuropsychological Testing and the Mentally Retarded</td>
<td>45</td>
</tr>
<tr>
<td>Summary</td>
<td>49</td>
</tr>
<tr>
<td>The Applications of Human Neuropsychology</td>
<td>50</td>
</tr>
<tr>
<td>Problems in Generalizability</td>
<td>50</td>
</tr>
<tr>
<td>Neuropsychology and Rehabilitation</td>
<td>52</td>
</tr>
<tr>
<td>Rehabilitation of the Mentally Retarded</td>
<td>54</td>
</tr>
<tr>
<td>Summary</td>
<td>57</td>
</tr>
<tr>
<td>Hypotheses</td>
<td>57</td>
</tr>
</tbody>
</table>

iv
<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>METHOD</td>
<td>61</td>
</tr>
<tr>
<td>Subjects</td>
<td>61</td>
</tr>
<tr>
<td>Materials</td>
<td>62</td>
</tr>
<tr>
<td>Procedure</td>
<td>69</td>
</tr>
<tr>
<td>RESULTS</td>
<td>71</td>
</tr>
<tr>
<td>DISCUSSION</td>
<td>87</td>
</tr>
<tr>
<td>Research Interpretation</td>
<td>87</td>
</tr>
<tr>
<td>Clinical Interpretation</td>
<td>97</td>
</tr>
<tr>
<td>SUMMARY</td>
<td>100</td>
</tr>
<tr>
<td>REFERENCES</td>
<td>102</td>
</tr>
</tbody>
</table>
## LIST OF TABLES

<table>
<thead>
<tr>
<th>Table</th>
<th>Description</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Means and Standard Deviations for Age and WAIS IQ for Each Age Group</td>
<td>63</td>
</tr>
<tr>
<td>2.</td>
<td>Comparison of Luria Corrected T-Scores for Older and Younger Groups</td>
<td>72</td>
</tr>
<tr>
<td>3.</td>
<td>Comparison of Luria Uncorrected T-Scores for Older and Younger Groups</td>
<td>75</td>
</tr>
<tr>
<td>4.</td>
<td>Means and Standard Deviations for AAMD Subscale and Total Percentile Scores for Total Group and for Age Groups</td>
<td>77</td>
</tr>
<tr>
<td>5.</td>
<td>Pearson Correlations Between AAMD Total Score and Luria Subscale Scores for Total Group and for Age Groups</td>
<td>79</td>
</tr>
<tr>
<td>6.</td>
<td>Discriminant Function Analysis of Luria and AAMD Scores</td>
<td>81</td>
</tr>
<tr>
<td>7.</td>
<td>Comparison of Uncorrected Left Hemisphere with Right Hemisphere Scale, and Verbal IQ with Performance IQ for Age Groups</td>
<td>84</td>
</tr>
<tr>
<td>8.</td>
<td>Comparison of Uncorrected Receptive Language with Expressive Language Scale for Age Groups</td>
<td>86</td>
</tr>
</tbody>
</table>
# LIST OF FIGURES

<table>
<thead>
<tr>
<th>Figure</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Corrected $T$-Score Profiles for Age Groups</td>
<td>73</td>
</tr>
<tr>
<td>2. Uncorrected $T$-Score Profiles for Age Groups</td>
<td>76</td>
</tr>
<tr>
<td>3. Corrected and Uncorrected $T$-Score Profiles for Total Group</td>
<td>83</td>
</tr>
</tbody>
</table>
INTRODUCTION

The meaning of the process of development has undergone change in recent years. In the past development was somehow a process meant only for children and adolescents, and it ended somewhere around age eighteen. It is now recognized that adults also undergo stages of change and growth which extend throughout their life span. Investigators have recently begun to define normal stages of adult development as well as aberrations from these norms.

It was a concern to understand a form of pathological adult development which launched the present study. Staff who worked in a long-term residential facility for the adult mentally handicapped observed abnormalities in the life span changes of residents afflicted with Downs syndrome, a specific form of mental retardation. Questions concerning the quality of these developmental changes and whether the pathological process could be detected early and thus counteracted became the core problem to be addressed.

In searching for answers to these questions it also became apparent that there is a lack of research data
specifically on adult Downs syndrome persons. Most re-
search was based either on children with Downs syndrome
or did not differentiate Downs syndrome from the numerous
other forms of mental retardation. The research which
separated adult mentally handicapped subjects into dis-
crete diagnostic groups was found to be a rare entity.
Psychologists who for many years had devoted effort to the
development of clear diagnostic categories and methods of
accurate diagnosis had neglected to understand that mental
retardation is only a global diagnostic term, much like the
term psychosis. Under the label of mental retardation are
subsumed numerous groups with differing traits, mental
capacities and physical abilities. Yet research had con-
tributed little toward understanding the characteristic
behavioral, cognitive and emotional differences among
these groups. Thus, the second problem to be addressed in
the study was to gain characteristic diagnostic data con-
cerning the subgroup of mental retardation which is Downs
syndrome.

Research on Downs syndrome has provided an under-
standing that such persons are afflicted with a genetic
abnormality which often results in intellectual subnor-
mality, language disorders, neurological and physiological
deviations and unique personality characteristics. These
descriptions provide a basic understanding of the nature
of Downs syndrome. What has been lacking in such data, however, has been a means of conceptualizing all these characteristics based upon a comprehension of the brain functions of a Downs syndrome person. Although it has long been recognized both that the brain is the organ of adaptive behavior and that the brain of a Downs syndrome individual is abnormal, few attempts have been made to integrate these ideas with descriptions of brain-behavior relationships for Downs syndrome. One major reason for this has been that the theoretical groundwork for such an integration has been unavailable to clinical psychology. Neurologists and brain specialists in the medical field have theorized about the mechanisms of the brain. Until the branch of psychology known as neuropsychology developed, however, there was little impetus for clinical psychologists to integrate their numerous observations about the behavior of humans with theories about the functioning of the human brain.

Scientists who have investigated schizophrenia have been the most successful in making this kind of theoretical leap. Knowledge about the cognitive disorder and biochemical reactions in schizophrenics has supplemented understanding of the affective nature of the disorder. The result of this integration has been a more complex, yet comprehensive appreciation of the nature of schizophrenia.
It would appear that the same process would enhance the study of mental retardation, and specifically Downs syndrome. This process would, however, require a major shift in orientation from current methods of diagnosis of mental retardation and its subgroups. The core of diagnostic and prescriptive analysis would become the conceptualization of brain-behavior relationships. This would suggest a devaluation of concepts such as the IQ, and descriptions of diverse deficits such as language and neurological deficits as the keys to complete diagnosis. Although honored in the past, these keys have proven limited in their ability to unlock as comprehensive an understanding of the mental retardation disorder as would an approach through neuropsychology.

The neuropsychological approach to diagnosis and treatment, especially in mental retardation, is a fledgling venture, and thus difficult to assimilate. It becomes necessary, then, to review past methods of understanding a disorder before exploring the neuropsychological approach and its applications. It is the goal of the subsequent discussion to assist in the development of a neuropsychological exploration of Downs syndrome by reviewing the current research on the nature of the disease, the development of the field of neuropsychology and finally, the applications of human neuropsychology.
The Nature of Downs Syndrome

**Discovery and incidence.** It has been over one hundred years since the first step in identification and research with a distinct group of mentally handicapped individuals emerged. In 1866 Langdon Down, a London physician, identified this group as characterized by noticeable physical features and mental inferiority. He attributed the cause to racial deviation. In the subsequent ninety years the theory of racial origins for the disease was discredited by observations that it occurred in all races and across all classes of economics and intelligence. Subsequent theories regarding its cause included factors such as advanced maternal age, abnormal conditions of the father and/or mother (alcoholism, syphilis, tuberculosis, psychopathology), a biochemical deficiency in the patient's organism, and preconceptual genetic alterations. It was not until 1959 that Lejeune, Gautier, and Turpin (1959) demonstrated that the cell tissue of these children contained forty-seven, rather than the normal forty-six chromosomes (Waranky, 1975).

At present the exact cause of the genetic abnor-
mality is not known. There appears to be some relationship to maternal age, with the incidence increasing past the age of forty years for women. Abroms and Bennett (1980) have suggested that paternal age may also be a more potent factor in bringing about the genetic deformity than was previously supposed. The incidence in the general population is currently reported to be one in seven hundred live births (Trunca, 1980).

Downs syndrome has been a well researched abnormality because it was the first known example of human aneuploidy (Crome & Stern, 1972). It is now known that in almost all patients with Downs syndrome, all or some of the body cells possess the extra genetic material. The most common (92 percent) is the Trisomy-21 type, which appears to occur because of faulty chromosome 21 separation during ovum formation. The second type, accounting for approximately 5 percent of the Downs syndrome population, results from a translocation from a chromosome 21 to either a member of the D or G group of chromosomes. The remaining 3 percent of Downs syndrome persons are called mosaics and possess two cell lines, one normal and one with the extra chromosome. In the mosaic type, the level of impairment is related to the percentage of cells affected by the extra chromosome (Donnell, Alfi, Rublee, & Koch, 1975; Shipe, Reisman, Chung, Darnell, & Kelly, 1968).
**Intellectual subnormality.** The most obvious result of the genetic abnormality is the intellectual subnormality which is consistently present in Trisomy-21 and translocation types and often present in mosaic types. It is now common practice, in fact, for these persons to be identified at birth, from their distinctive physical characteristics and labeled mentally retarded immediately. The rate of intellectual development in these children is known to be slower than normal. Dicks-Mireaux (1972) administered the Gessell Scales five times to Downs syndrome infants up to the age of seventy-eight weeks. He concluded that the abnormal rate of development appears by the age of sixteen weeks, and that from that age onward there is a progressive deterioration in the rate of development. Others have concurred that the intellectual deterioration does appear soon after birth. Kirman (1973) concluded that it is unclear, however, whether this inferiority is due to a cerebral process which becomes worse as the child grows, or, more likely, to a manifestation of a fixed anatomical defect which simply appears at an early age.

The intellectual capacities of Downs syndrome individuals have been compared with those of other groups of mentally retarded persons. Johnson and Olley (1971) compared the Mental Ages (MA) of Downs syndrome, non-organic retarded and organic retarded groups. They found
the Downs syndrome MA's to be lower than those of the non-organic group, but not significantly different from the brain damaged group. Mentally retarded children as a whole have been reported to obtain higher Performance IQ than Verbal IQ on the WISC (Sattler, 1974), and similar results on the Wechsler-Bellevue have been reported for mentally retarded adults (Matarazzo, 1972). It is suspected that Downs syndrome children and adults would produce a similar Verbal-Performance IQ pattern, although no past studies have been done to examine this specific issue.

For many years the intellectual defect of Downs syndrome children had been considered so severe throughout life that they were labeled incapable of progressing or benefitting from most types of intellectual training and stimulation. However, recent research and experience have questioned that conclusion. Connolly (1978), after testing one hundred eighty Downs syndrome children on the Stanford-Binet and WISC, concluded that there is not a homogeneity of mental abilities in the group. Some subjects showed evidence of the effects of immature or faulty cerebral development, while others showed evidence of cerebral assault as well. He found that higher functioning Downs syndrome persons were not so rare as was previously suspected, and attributed this to the possible
results of the current trend toward home rearing and early education for these children. In reviewing the literature on IQ data, Baroff (1974) concluded that an average IQ between forty and forty-five was found for many preschool aged Downs syndrome children. However, like Connolly, Baroff noted that home rearing appeared to raise IQ scores from five to ten points for this age group. Rynders, Spiker, and Horrobin (1978) have stressed the importance of this last point. They emphasized that the major methodological problem in research with Downs syndrome persons is the lack of control of residence (home vs. institution) as well as such variables as sex, age and selection criteria. Thus general conclusions regarding the intelligence levels and limits of educability of Downs syndrome children may be faulty.

Language disorders. As with the literature on intellectual subnormality, the research on language disorders in Downs syndrome does not always distinguish this group from other groups of mentally retarded individuals. Additionally, the relationship between level of intelligence and level of language development is an old argument which has received considerable attention in the general literature on mental retardation. Theorists tend to disagree on the importance of language development to the development of cognitive processes.
The literature reveals that the incidence of language disorders in all mentally handicapped groups is high. Spreen (1965a), in reviewing the research studies on correlations between language dysfunction and intelligence, suggested that the two abilities may be somewhat independent. While many studies supported an inverse relationship between intelligence and the incidence of language disorder, closer analysis revealed these only represented a moderate range of correlations (-.31 to -.41). Furthermore, incidence of language problems varied not only with level of overall intellectual impairment, but also with the type of assessment, i.e., the specific components of language which were assessed.

Spreen concluded that the incidence of language impairment ranged from almost 100 percent for the lowest functioning mentally retarded persons (IQ's below 20), to about 45 percent for the mildly retarded group (IQ's between 50 and 69). Specific components of language, such as the age of onset of talking, speech sound development and acquisition of phonemes showed a consistent, but low correlation with standard IQ tests. On the other hand, relatively high correlations emerged between measures of IQ and vocabulary up to about the MA of seven to eight years. A moderate correlation of speech productivity (number of words spoken during a standard situation) with
both IQ level and vocabulary level was obtained. Some studies reviewed by Spreen found a small lag in such measures as sentence length, sentence complexity, discrimination of speech sounds and noun percentages when mentally retarded subjects were compared to MA matched non-mentally retarded subjects, but other studies did not report such differences. Finally, Spreen found that in some studies, in which simple verbal measures such as vocabulary size were used, mentally retarded subjects were superior to normals of equal MA levels.

In his review of language research with mentally retarded subjects, Spreen (1965a) found few inconsistencies in comparing Downs syndrome with non-Downs syndrome mentally retarded persons. The inconsistencies which he did report were that noun percentages, which represent concrete level of speech, dropped at a later age with Downs syndrome subjects, and there was a higher percentage of articulation disorders, voice defects and stuttering in the Downs syndrome subjects. He also reported differences in verbal MA levels between Downs syndrome and non-Downs syndrome subjects in institutions, but was unable to find the same discrepancy in studies conducted in day schools.

Cornwell (1974) proposed that the limitation in verbal skills in Downs syndrome subjects is due to the
disparity between receptive and performance verbal abilities, with receptive generally being higher. In his investigation, some Downs syndrome subjects could demonstrate their understanding of an object's functioning through gestures, mimicry and peripheral verbalizations, but could not convey their understanding through verbal expression. Cornwell concluded that "it is evident, then, that poor language expression can often mask the actual level of verbal functioning in children with Downs syndrome, so that conventional criteria are inadequate to assess fully their verbal capacities" (p. 187). Deficiencies in articulation can only thus account in part for deficiencies in verbal abilities. Cornwell suggested that higher brain functioning must be analyzed to explain fully the phenomenon.

The argument concerning the relationship between language level and the development of intellectual processes is important to the understanding of mental retardation. If the relationship is presumed to be a strong one, then level of cognition could be inferred directly through assessment of language. If, however, the relationship is not considered so strong, then language ability and intellectual processes need to be assessed separately. The notion that the relationship is strong is exemplified in the work of Luria, while the opposing view
is taken by researchers who have examined intellectual development in persons with limited verbal skills, particularly the deaf.

Luria emphasized the relationship between the development of language functions and intellectual processes and concluded that language is the instrument of thinking. He differentiated levels of mental deficiency based not on IQ, which he considered subject to class biases, but on "the extent of the under-development of the system of verbal connections and the degree of the disturbance of their participation in the regulation of activity" (Luria, 1963, p. 197). In reaching these conclusions Luria examined the works of his predecessor, L. S. Vigotsky, who felt that the road to the examination of higher mental processes was through examination of the reorganization of mental processes that comes with speech. As early as 1929 Vigotsky had observed that when a four to five year old child is faced with a difficult problem, external speech, or verbal mediation, arises (Luria & Yudovich, 1968). For Luria, the development of motor behavior, which he termed the first signalling system, is followed by the development of language as the second signalling system. As the child grows, the language and motor connections alter, so that by age six spoken language becomes the "inner language" of action. "The specific practical activity begins
to be mediated by speech and is connected into that system of internal operations which is the most important acquisition in a child's normal mental development" (Luria, 1963, p. 7).

For Luria, then, mental retardation was essentially a dissociation between the interaction of the two signalling systems, verbal and motor behavior, with the regulation of behavior by speech being faulty. In the mentally retarded child activities remain at the level of specific external actions and there is a delay in the formation of internal mental processes which is related to the under-development of speech (Luria, 1963).

To confirm these conclusions Luria and Yudovich (1968) studied a pair of monozygotic twins, both delayed in speech development and also further delayed by the fact that they did not need language because they had developed an ability to understand each other, a phenomenon the authors termed the "twin situation." At pre-test both twins had little language and what language they used was highly interlocked with actions. The boys also could not organize complex play. Luria and Yudovich separated the twins into different pre-school classrooms and provided speech therapy to one twin. Both twins improved in speech, presumably due to the reduction of the "twin situation," but the trained twin surpassed his brother in intellectual
development as well, as exemplified by his ability to de-
code stories, make up games and create new meanings, re-
late stories, develop judgment, detect absurdities in
drawings, and classify objects beyond the limits of visual
perceptions.

Others, however, have disputed Luria's heavy empha-
sis upon the development of language functions as critical
to intellectual development. Spreen (1965b) cited re-
search with deaf persons, who do not verbally mediate, yet
develop concepts, to support his questioning of Luria's
theory. Zigler and Balla (1971) specifically cited re-
search in which deaf children and retarded children were
found to be superior to non-retarded children on a task
measuring understanding of the concept of symmetry. These
authors, after reviewing research relevant to Luria's
theory, concluded that there is not any convincing evidence
to support Luria. Their conclusion was based upon several
factors: ambiguity regarding Luria's concepts, methodolog-
ical problems (inconsistent criterion measures and tasks
employed to measure the underlying variables), contra-
dictory results, and possible confounding by motivational
factors for the retarded group.

In general, it can be said that Downs syndrome per-
sons possess many of the language impairments that other
mentally retarded groups display. Only a few discrepan-
cies have been reported to distinguish Downs syndrome from other groups. One of these may be the greater development of receptive language over expressive language for Downs syndrome subjects, as suggested by Cornwell (1974). The literature also does not clarify the strength of the relationship between the development of language skills and intellectual processes for those who have language impairments. It appears possible, from the moderate correlations reported by Spreen (1965a) and the arguments of Zigler and Balla (1971) that Luria's conclusion that language is the instrument of thought may be faulty.

Physical and neurological abnormalities. Not only do Downs syndrome subjects have intellectual and language deficits, but physiological and neurological deficits as well. The physical character of the disorder was the first means of identifying afflicted persons. Perhaps because the physical aspects of the syndrome are so evident, much effort has been placed toward understanding these physical problems and attempts to cure them through medical means. As the discovery of chlorpromazine provided alleviation of some of the major symptoms of schizophrenia, it was hoped that a similar medical intervention could bring Downs syndrome persons closer to normality. These efforts have, however, been generally unsuccessful.
Physically, Downs syndrome persons have smaller than normal birth weights (Pueschel, Rothman, & Ogilby, 1976); they also have less than normal brain weights, but generally greater brain weights than other mentally retarded populations. Some neuropathology studies have revealed that many Downs syndrome persons have roundish shaped brains, with small frontal lobes, brainstems and cerebellums, and a narrow superior temporal convolution (Crome & Stern, 1972). Others suggest, however, that there is little agreement over neuropathological findings on the size of Downs syndrome brains (Coleman, 1973). The incidence of physical stigmata is high in Downs syndrome, but little correlation has been found between degree and number of physical stigmata and level of intellectual functioning (Dunsdon, Carter, & Huntley, 1960; Shipe et al., 1968). Rundle and Sylvester (1978) found that biological mechanisms responsible for the regulation of growth in Downs syndrome are similar to those of normal children. They also found that sex-dependent growth factors in normals function similarly in prepubital and adolescent growth of Downs syndrome subjects. However, hyperthyroidism was more frequent among older Downs syndrome persons than normals and may account for their generally shorter stature (Rundle & Sylvester, 1978).

The abnormal Downs syndrome brain has been examined
through use of the electroencephalogram (EEG), with the hope that consistent patterns would emerge to differentiate the Downs syndrome person from normals and other mentally retarded groups. Goldie, Curtis, Swendsen, and Robertson (1968) compared the sleeping EEG's of normal and Downs syndrome newborn infants. They found that the Downs syndrome newborns had longer average duration of non-REM sleep, increased period of transition from REM to non-REM sleep, and more frequent body movements, especially side-to-side head movements. They also recorded the absence of EEG patterns which are usual in non-REM sleep of normal infants. Deliberate awakening in non-REM sleep was found to be more difficult in some of the Downs infants than in normals, but was not accompanied by the usual irritable state. Kirman (1973) studied the EEG patterns of twelve month old infants and reported that Downs syndrome subjects were unable to extinguish central responses to auditory stimuli which were not reinforced, while normals were able to do so. Ellingson and Lathrop (1973) investigated higher alpha frequencies, as these may suggest more rapid information processing and therefore higher intelligence. They discovered no significant differences among alpha rhythms of Downs syndrome, psychiatric and normal college student subject groups.

Ellingson, Menoloscino, and Eisen (1970) reviewed
the EEG studies on Downs syndrome. They concluded that there is a wide variety of EEG abnormalities which do not correlate well with behavioral and neurological signs. They also concluded that there is a greater frequency of EEG abnormalities among Downs syndrome than normals, but a lower frequency of such signs for Downs syndrome than for non-Downs syndrome mentally retarded groups. Fewer Downs syndrome children than non-Downs syndrome retarded children have epilepsy (a neurological disease often related to EEG abnormalities), but the incidence of epilepsy also increases with age in Downs syndrome (Veall, 1974).

Drug research has also been a major area of investigation in the physiological studies of Downs syndrome subjects, although generally with inconclusive results. Drugs have been utilized to produce a variety of effects with the mentally retarded population in general. The history of drug use with mentally retarded persons dates back to the use of sedatives, hypnotics, stimulants and anticonvulsants in the 1930s, use of antihistamines in the World War II era, and use of minor and major tranquilizers in the 1950s. Such drugs were used in the past not as a cure for mental retardation, but more as chemical controls when mentally retarded persons displayed psychotic symptoms or behaviors such as aggression and agitation (Share, 1976).
By far the most hopeful attempt in this area has been the research with 5-hydroxytryptophan (5-HP T), an amino acid which is converted to serotonin, a substance known to have a potent effect upon brain metabolism. Alterations in serotonin activity have been confirmed in phenylketonuria (PKU) patients, and early dietetic interventions have proven effective in reducing the level of mental retardation in PKU infants (Coleman & Hur, 1973). In Downs syndrome, low levels of serotonin have also been found (Tu & Zellweger, 1965). Attempts to administer 5-HP T to Downs syndrome subjects to counteract the low serotonin levels have not led to promising results. In one study (Kirman, 1973) the 5-HP T was being excreted in the urine of the subjects without altering the serotonin levels or neurological status of the subjects. When tryptophan, a version of the drug, was administered, the level of 5-HP T was increased in the cerebrospinal fluid but still not to a normal level.

Mary Coleman (1972; 1973) has been the major proponent of the effects of the serotonin research in Downs syndrome. She initially suspected that the administration of 5-HP T would result in improved speech development and muscle tone and hypothesized that brain mechanisms could also be affected. In her first, uncontrolled study Coleman reported that the administration of 5-HP T to
Downs syndrome infants appeared to affect muscle tone, age of walking, incidence of cardiac disease, strabismus, tongue protrusion, alertness and activity level, height, weight and head size. However, in a three-year, controlled, double blind study, she reported only one short-term effect, the evidence of increased muscle tone up to four months of age. After that age, 5-HTP had a deleterious effect until the age of three years, after which no effect could be discerned. Increased activity level of 5-HTP subjects was partially confirmed, but Coleman herself questioned the possible role of staff interactions to help parents create a positive environment for their children upon such results. No effect of 5-HTP on overall mental development, as measured by the Bayley Mental Development Scales, was found. Additionally, serious side effects were reported, including significant increase in EEG abnormalities in the experimental group and development of infantile spasms. Coleman's final conclusion was that the administration of 5-HTP is not recommended (Coleman & Steinberg, 1973). Share (1976) commented that poor control, failure to show any long-term effects and possible side effects of the drugs limit the credibility of Coleman's work. Share further commented that such poor methodological control is typical of drug treatment studies in Downs syndrome. He emphasized the placebo effects of staff and parental attention, noted by Coleman
herself, as particularly troublesome to such research. Thus, the drug research and EEG studies have been unsuccessful in fully characterizing or altering the brain functioning of Downs syndrome persons.

One other neurophysiological finding has further complicated the puzzle, as it is a cerebral condition which occurs later in the adult life of the Downs syndrome individual. It has been concluded from neuropathological studies that not only is cerebral development in Downs syndrome abnormal in childhood, but also the cerebral aging process may be abnormal. The findings indicate that there is an earlier than normal onset of several neurophysiological and behavioral signs of senility similar to the process known as Alzheimer's disease. Results such as these have been rarely found in other mentally retarded groups. Jervis (1948), when performing autopsies on three Downs syndrome cases, aged 47, 42 and 37, all of whom had experienced personality changes and mental deterioration previous to death, discovered senile plaques and Alzheimer's changes in the brains of all three subjects. Several studies have since confirmed these findings, although in various brain locations (Burger & Vogel, 1973; Callner, Dustman, Madsen, Schenkenberg, & Beck, 1978; Ellis, McCulloch, & Conley, 1974; Jervis, 1970; Malamud, 1972; Ohara, 1972; Olson & Shaw, 1969; Solitaire & Lamarche, 1966).
Alzheimer's disease is a genetically determined disorder. It has been known to appear in juvenile normals, although rarely. Alzheimer's disease accounts for 47 percent of the diagnosed cases of dementia in the general population and thus is the largest diagnosable cause of dementia (Wells, 1979). There are three stages in the development of the disease. The first stage results in amnesia, depression, distress over handicaps, loss of efficiency and orderliness, superficial conversation and a flippant attitude. In the second stage symptoms of disorientation, increased depression, dysarthria, confusion and decreased performance appear, while the third and final pre-death stage is characterized by Parkinsonian symptoms, incontinence, epilepsy, apraxia and possible delusions and hallucinations. Current research suggests that such changes are more likely to appear in Downs syndrome subjects after age thirty-five with minimal neuropathological findings occurring before age thirty (Owens, Dawson, & Losin, 1971). Malamud (1972) has reported 100 percent incidence of such changes in Downs syndrome persons above age forty on whom autopsies have been performed. This is considerably earlier than the cerebral aging process begins in non-Downs syndrome persons of both normal and subnormal intelligence.

The early detection of this unusual aging process
is not easy, however. In the general population, many early signs of the disease suggest a functional disorder and initially result in the patient being brought to a psychiatrist for treatment. Fully developed senile dementia is often only recognized as an organic disease once it has reached the last stages (Wells, 1979). Similar problems affect the early diagnosis of the disease in the Downs syndrome population. Owens et al. (1971) reported that only occasionally do the dementia and psychiatric problems occur in older Downs syndrome subjects, and Neumann (1967) suggested that when such personality changes do occur, the disease is past its early stages. Similarly, a progressive and nonspecific slow wave EEG disorganization is associated only with the later stages of the disease and thus the EEG is not helpful in early detection (Crapper, Dalton, Skopitz, Scott, & Hachinski, 1975). Some Downs syndrome patients display typical signs of slow mental deterioration, but the majority show no clinical signs. When symptoms do occur, they are inconsistent, and the variety reported include such diverse signs as visual agnosia, pathological reflexes, incoordination and extremity tremors, and increased incidence of seizures. Crapper et al. (1975) have suggested the presence of a paradox which hampers early detection. Since Downs syndrome subjects are presumed to already possess a reduced number of neurons, the presence of
Alzheimer's should result in a nervous system which is very sensitive to neuron loss and thus clinical signs should appear early in the course of degeneration. This does not occur, however. Ellis et al. (1974) have suggested that the detection of further deterioration in the face of long-standing mental deficiency may be very difficult.

**Personality abnormalities.** Contributing to the difficulty in detecting personality changes which may reflect brain alterations in Downs syndrome is the fact that there exists little baseline personality data on the Downs syndrome adult. Over the years, there has developed a stereotyped notion, which is unverified empirically, to describe the personality of Downs syndrome persons. As children, they are expected to be playful, affectionate and passive and these traits are often expected of the Downs syndrome adult as well. Psychiatric disturbances and behavioral problems are popularly assumed to be low in frequency, because of the presumed docility.

Personality testing to confirm such theories has been difficult with this population because of the subjects' cognitive limitations which hamper the valid use of personality tests. Nevertheless, some studies have examined the validity of the notion of the universally passive and docile personality of all Downs syndrome
persons. Sarason and Doris (1969) observed that the incidence of psychiatric disturbances is lower in Downs syndrome persons than in other forms of mental retardation. Rutter (1971) suggested that the lower frequency of such difficulties when compared to other mentally handicapped groups may be related to lower frequency of epilepsy. Menolascino (1965) reported that the incidence of psychiatric problems in Downs syndrome children up to age eight years is comparable to the incidence of such problems in the general population of children of the same age.

Brink and Grundlingh (1976) compared the Rorschachs and Human Figure Drawings of Downs syndrome and non-Downs syndrome retarded adults to differentiate the two groups on personality variables. On the Rorschach the Downs syndrome group revealed a better developed self-concept and higher level of social maturity and awareness, but these results were not confirmed by the Human Figure Drawing data. The authors suggested that there is a need for more studies of this type. They also indicated that such studies should utilize testing techniques which rely more upon visual perception skills than verbal skills in order to obtain valid analyses of personality traits in Downs syndrome persons.

These few studies reveal that there is very little known about the typical personality characteristics of the
Downs syndrome adult. If personality changes may signify brain changes, it would be helpful to have baseline personality data available. However, the state of the art is so primitive at this time that the measurement of personality variables would be useless in revealing information on brain functioning.

**Summary.** It was stated earlier that the literature lacks both data on the Downs syndrome adult and a unifying theory on brain-behavior relationships for such subjects. As the above discussion exemplifies, the literature does provide a series of related but unconnected pieces. These pieces succeed in describing the various deficits related to the disorder, but fail to provide an integrated picture of how the Downs syndrome brain functions to produce the deficits.

However, a neuropsychological approach to understanding Downs syndrome may provide the needed concepts. It is the task of neuropsychology to tie observed behaviors to theories of how the brain operates. This should lead in turn to the testing of specific hypotheses about the nature of Downs syndrome. Neuropsychology may thus be able to provide a more appropriate and useful method of examining diverse phenomena such as Performance IQ greater than Verbal IQ (an intellectual deficit), receptive language vs. expressive language differences (a language
deficit) and the early cerebral aging process (neurological abnormality) in Downs syndrome. The results of such research may then suggest particular treatment strategies or may provide explanations for the success or failure of current strategies. The first step in this process of understanding Downs syndrome is to determine the usefulness of neuropsychological testing methods to achieve the goals of improved diagnosis and treatment.

Human Neuropsychological Testing

History of human neuropsychological theory. The study of brain-behavior relationships has origins in ancient history. Pythagoras believed that human reasoning originated in the brain and Hippocrates was the first to recognize that an injury to one side of the brain resulted in paralysis on the opposite side of the body. In the thirteenth century Albertus Magnus concluded that the frontal lobes, cerebellum and ventricles were all responsible for behavior, and in the seventeenth century Descartes located mental abilities in the pineal gland (Golden, 1978).

Localizationist theories became prominent after 1861, when Broca located motor speech in the left posterior frontal lobe and Wernicke soon afterwards located speech understanding in the posterior temporal lobe. Opponents of the localizationist theory proposed the equipotential
theory, which suggested that the whole brain is involved in most mental processes. Thus the extent of destruction of the brain, rather than just the location, was considered critical to the loss of a specific function. Proponents of this theory were Flourens, Goldstein and Lashley (Golden, 1978).

During the second half of the nineteenth century J. Hughlings Jackson proposed an alternative theory which has been furthered most recently by the Russian neuropsychologist, A. R. Luria. Jackson suggested that more complex mental abilities, such as speech, are made of a combination of less complex skills. If any of these simpler skills is impaired, the higher mental function may also be impaired. Jackson's ideas negated the notion that any particular higher mental ability is located in one particular area within the brain. A higher mental ability is, instead, a result of complex interactions among several areas of the brain. His conclusions in this area grew out of observations that precise, localized lesions never caused the breakdown of a complete mental function. He proposed a system of low, medium and high levels of organization within the brain, with these three levels further interacting to produce complex mental functions (Golden, 1978; Luria, 1979).
The field of neuropsychology had its birth in the debate between the localizationist theorists and the equi-potential theorists, with most modern neuropsychologists accepting a theory more akin to Jackson's complex structural theory. The field of neuropsychology achieved greater impetus toward development during World War II, when psychologists were being called upon to assess the effects of damage and rehabilitative potential of soldiers who received head wounds during the war. This practical need led to the further development of objective methods for measuring brain-behavior relationships.

Boll (1977) has recently summarized the current role of neuropsychology, and his concepts support the notion that a neuropsychological investigation of Downs syndrome is beneficial. Boll proposes that the purpose of neuropsychology is not primarily to focus upon pathological processes in the brain, but is to understand human ability and to evaluate an individual's strengths and weaknesses across many abilities. The Downs syndrome individual may present his own characteristic pattern of such strengths and weaknesses. Neuropsychology is unique in that, perhaps more than in any other area of psychological assessment, the information gained from diagnostic procedures is directly linked to methods of remediation. This direct link could lead to new methods of
training persons with disorders such as Downs syndrome. Furthermore, Boll suggests that neuropsychological procedures thusfar developed have proven sensitive to even subtle changes in brain-behavior relationships and thus pick up such changes in a way that some medical tests cannot do. Boll concludes that neuropsychology has a vitally important role in the understanding of human behavior, which is the task of all psychology: "Despite the importance of many external factors, it remains true that the brain is the integrating or central organ of adaptive behavior" (Boll, 1977, p. 65). If the brain can be better understood, so also can human behavior.

While few would dispute Boll's general statements of goals for neuropsychology, clinicians in the field do dispute the approaches and methods utilized to achieve the goals. Two major approaches have been adopted. On one hand, American neuropsychologists have emphasized a quantitative, practical approach which lacks a unified theory of how the brain functions. On the other, Soviet neuropsychologists have advocated a qualitative, non-statistical approach. Out of the former grew the Halstead-Reitan Neuropsychological Battery, while the latter was advanced by the work of A. R. Luria.
The Halstead-Reitan approach. The first neuropsychological test to be published was that of Ward C. Halstead, who established the first laboratory to study human neuropsychology at the University of Chicago in 1935. His perhaps optimistic intent was to study what he called "biological intelligence," a form of cognition which is free of cultural influences and relates primarily to the human nervous system. From his laboratory Halstead developed a battery of tests to distinguish those patients with cerebral lesions from those without such lesions. The results of these tests were summarized in one score, called the Impairment Index. Utilizing a specific cutting score for each test and an overall cutting score of five out of ten tests falling within the abnormal range, Halstead was successful in predicting brain damage in general and frontal lobe brain damage in particular. His norms for the cutting scores were obtained from a population of thirty normal subjects aged fourteen to fifty years (Lezak, 1976).

The original Halstead test was revised and expanded by Ralph M. Reitan. The battery in its present form consists of most of Halstead's original tests, the WAIS, MMPI, Trail Making Test, an aphasia examination and tests of sensory-perceptual functions, and is known as the Halstead-Reitan Neuropsychological Test Battery. It con-
continues to be the most widely used neuropsychological test battery today (Reitan, 1966).

The validity and reliability of the Halstead-Reitan Battery have been examined over the past twenty years (Boll, 1978; Klove, 1974). In 1955 Reitan compared fifty pairs of brain damaged and non-brain damaged subjects on the Halstead tests and found significant differences for all tests except one, which was then dropped from the battery (Reitan, 1955). Vega and Parsons (1967) cross-validated and obtained similar results regarding the battery's sensitivity to organicity. Filskov and Goldstein (1974) compared the battery's ability to discriminate brain damaged from non-brain damaged individuals with the ability of commonly used medical tests (brain scan, flow studies, EEG, angiogram, pneumoencephalography, skull x-rays) to do the same. They found that in many cases the hit rate for the neuropsychological battery was superior to that of the medical tests. Matarazzo, Matarazzo, Wiens, Gallo, and Klonoff (1976) reported a high degree of clinical and psychometric reliability for the Halstead-Reitan for four major groups of subjects: normals, older cerebrovascular subjects, chronic schizophrenics and carotid endarterectomy patients. There have been numerous other studies which have confirmed the reliability and validity of the Halstead-Reitan battery and its sensitivity to the presence
and location of brain lesions (Boll, 1978; Klove, 1974).

The result from Halstead's initial work has been a highly standardized and reliable approach to neuropsychological testing that is capable of quantification and mathematical analysis. The approach is also very successful in making dichotomous decisions regarding presence or absence of brain damage, location of injury within the two cerebral hemispheres and the acuteness or chronicity of the injury. In spite of the high level of acceptance of this approach, it has been criticized for its inappropriateness for patients with sensory or motor handicaps, as such conditions sometimes require the abandonment of large portions of the test. The full Halstead-Reitan battery also takes large amounts of time to administer (often up to eight hours per patient), cannot be administered easily at bedside, and involves the use of very expensive equipment, all of which inhibit its widespread use (Luria & Majovski, 1977). However, through a recent unique marriage of Soviet and American neuropsychological approaches, critics of the Halstead-Reitan are suggesting an alternative based on the theoretical orientation of Luria. This alternative also has advantages for those interested in the understanding of mental retardation.

The Luria approach. Alexander Romanovich Luria (1902-1977) was influenced early in his career by Vigotsky,
who was interested in creating an experimental psychology of higher psychological functions. Together Vigotsky and Luria investigated the development of higher cognitive processes in children, particularly the interplay of motor and speech in young children. After Vigotsky's death in 1934, Luria pursued a career in medicine and specialized in neurology. World War II brought him the opportunity to study head injured patients and to expand further his own ideas on brain-behavior relationships. He also examined developmental cerebral abnormalities associated with the mentally handicapped. The last twenty-five years of his life were devoted to the expansion of his own complex theory of human neuropsychology (Luria, 1979).

Early in his neurological studies Luria rejected the localizationist theory as too simplistic. Like Jackson, Luria's own observations of patients with very localized lesions revealed that the result was not the loss of one function, but sometimes the loss of many heterogeneous functions (Luria, 1965). He also believed, like Jackson, that the brain functions as a complex system of interrelated parts.

Luria turned away from the use of American style IQ tests as a way of assessing brain-behavior interactions. Concerning his study of relative influences of heredity
Luria commented:

We were particularly unhappy with the use of standardized IQ tests as the indicators of intellectual development. These tests, which were developed on a purely pragmatic basis to predict school performance, seemed to us then, as now, to be a hopelessly atheoretical and opaque means of observing the structure of higher psychological functions (Luria, 1979, p. 82).

Luria's implications here appear to be that the popular method of IQ testing was simplistic and inadequate for explaining brain functioning. Nevertheless, IQ testing methods continue to be one of the primary methods for defining mental retardation.

Luria adopted, instead, the idea of the organization of mental functions according to systems, which he termed "functional systems," and which cannot be simply localized in the brain. According to Luria, one functional system is not the result of the action of one specific area of the brain, but rather many areas of the brain interacting in various ways to produce various behaviors. Thus each area of the brain may participate in several functional systems. Luria spoke of the basic character of a functional system as consisting of "the presence of an invariant task, performed by variable mechanisms, which bring the process to a constant, invariable conclusion" (Luria, 1979, p. 124).
Luria stressed the flexibility of the functional systems. During childhood the roles played by different areas of the brain in organizing complex behaviors are different than for adults. For example, in children sensory areas are more important than they are for adults. Thus the systemic organization of the brain changes as the child grows (Luria, 1979). Similarly, the disruption of part of a functional system in an adult does not necessarily mean that the behavior involved is permanently lost, as nonaffected parts of the system may be utilized to reorganize the way in which the behavior is achieved. This concept of "re-wiring" is the key to rehabilitative processes (Luria, 1973). It also has implications for understanding mental retardation. The abnormal cerebral structure in Downs syndrome, for example, may indicate that learning occurs by use of different functional system pathways than in normals.

Luria described three principal functional units of the brain, each of which is further subdivided into three cortical zones. The first principal functional unit is that which is responsible for regulating the general tone and alertness level of the organism. An important structure of this unit is the reticular activating formation. It plays a role not only in the activation of the organism on metabolic and sensory input levels, but also
interacts with various parts of the brain to regulate consciousness, emotion and memory. The second principal functional unit is responsible for receiving stimuli, analyzing and storing information, and includes parts of the cortex responsible for visual (occipital lobe), auditory (temporal lobe) and general sensory (parietal lobe) functions. The third functional unit includes the pre-frontal and frontal portions of the cortex and is responsible for the forming of plans, regulation and unification of activity (Luria, 1973).

The further breakdown of each of the principal functional units reveals a hierarchical structure of primary, secondary and tertiary portions. The tertiary portions are the last to develop in children and represent the most complex form of mental organization. For example, the tertiary portion of the third principal functional unit operates as a superstructure overseeing the regulation of all behavior and is thus responsible for playing a more complicated role. In instances of mental retardation, it could be hypothesized that this complex tertiary superstructure is generally more impaired than are lower structures.

No one complex psychological process takes place without input from all three principal functional units, and it is the pattern of interaction of these various
areas of the brain which is responsible for a given behavior. For example, the process of perception requires the alertness of the organism, reception and analysis of the percept and the determination of any necessary action resulting from that analysis. A similar interaction could be posited to describe the complex behaviors known as reading, writing or speaking. Thus it can clearly be seen that Luria has gone far beyond the simple notion of precise localization of functions within the brain (Luria, 1973).

In order to confirm these hypotheses, Luria investigated the results of brain lesions in various areas of the brain to determine the contribution of that area to the organization of complex behavior. During his years of contact with brain injured patients Luria also developed his own methods of assessing the lesion's effects on the patient's behavior. His method of assessment consisted primarily of simple tasks which could be administered at bedside to patients, such as the imitation of simple motor movements, the detection of sensory input and basic writing and reading tasks. For Luria, this method of assessment was useful not only in determining the presence or absence of a specific behavior, but also in analyzing the quality of the patient's response. Also for Luria, who was unconcerned with quantitative approaches to assess-
ment, it was unnecessary to develop any standardized or scorable method of assessment. Thus for many years his diagnostic procedures remained unavailable to others. It was not until 1975, when Anna-Lise Christensen published a description of Luria's techniques and attempted to produce a manual for their administration, that Luria's methods became available for others to use (Christensen, 1975). Even Christensen's attempts, however, were criticized by proponents of a standardized and quantitative approach, for variance of procedure from patient to patient, depending upon the clinician's impressions found during an initial interview, and for the lack of any cross-validation data (Reitan, 1976). Thus American psychologists, who were proponents of the quantitative Halstead-Reitan type of approach to neuropsychological assessment initially rejected the Luria approach because of its lack of standardization.

**Standardization of the Luria approach.** In 1978 Charles J. Golden and his associates at the University of South Dakota published their first standardization of Luria's methods, and in the following year they issued a revised manual. During this period Golden moved his work to the University of Nebraska and entitled his published test the Luria-Nebraska Neuropsychological Battery (Golden, Hammeke, & Pruisch, 1979).
Besides making Luria's work more available to the public of neuropsychologists, Golden sought to develop a scale that met the demands of both flexibility and standardization. He also sought to develop a comprehensive test battery, one which would be able to isolate specific dysfunctions as well as evaluate all areas of complex behavior. The resulting scale consists of 268 individual items, each of which is considered of diagnostic significance in itself. The items are organized into subscales measuring complex functions. The complex processes assessed by the scale include general intelligence, memory, perceptual and perceptual-motor performances, language functions, sensory skills, reading, writing and mathematical skills. Finally, Golden sought to develop a scale that was inexpensive and less time consuming than the Halstead-Reitan and which could be administered at bedside, much like Luria himself would do (Golden, Hammeke, & Purische, 1978).

These particular advantages also make the Luria testing approach attractive to those interested in studying mental retardation. The simplicity of many of the items makes it easier for the mentally retarded person to respond and thus makes more data available. The discrete nature of the items makes it unnecessary to assume that later items will be failed because of failure on an earlier
item. Since mentally retarded subjects tend to fail even easy items on tests, it often becomes necessary to discontinue large sections of more traditional tests. On the Luria, the testing can often continue after failures because of the diverse nature of the items.

Validation and reliability investigations are still in process for the Luria-Nebraska Scale. Data which have been published thusfar support the scale's ability to discriminate brain damaged from non-brain damaged subjects, to lateralize lesions within the two cerebral hemispheres, and to localize to the four major quadrants of each hemisphere. Utilizing an experimental group with known neurological impairment and a control group including spinal cord injury patients, the Luria-Nebraska's subscale indices correctly classified 86 percent of the brain damaged group and 100 percent of the non-brain damaged group, with an overall hit rate of 93 percent (Hammeke, Golden, & Purisch, 1978). In a cross-validation study, Moses and Golden (1979) reported similar results. Overall discriminant analysis and t-test analyses of each item subscore revealed that of the unrevised original 285 items, 253 significantly discriminated between brain damaged and non-brain damaged groups (Golden et al., 1978). The test has also been found to obtain an overall hit rate of 88 percent in its ability to discriminate neurological
patients from chronic schizophrenic patients (Purisch, Golden, & Hammeke, 1978).

Attempts to lateralize (i.e., locate the injury within either the right or left cerebral hemisphere) from the Luria data have led to the development of empirical scales. Osmon, Golden, Purisch, Hammeke, and Blume (1979) correctly classified fifty-nine out of sixty patients with lateralized brain lesions. Their classifications were obtained by analyzing test items which compared functions on the two sides of the body and which measured skills known to be related to individual hemispheres. Subsequently, McKay and Golden (1979) empirically derived additional subscales to differentiate left hemisphere lesions from right hemisphere lesions. Further research with these empirical scales revealed a hit rate of 87.8 percent for left brain damaged patients and 86.6 percent for right brain damaged patients. Significant differences between brain damaged and non-brain damaged groups on the empirically derived left hemisphere and right hemisphere scales were also reported.

Few localization studies have been completed to date. One published study revealed that different patterns emerged on the test for patients with lesions in different locations in the brain (Lewis, Golden, Moses, Osmon, Purisch, & Hammeke, 1979). Golden (1979) also has
reported that he was able to discriminate patients with a specific disorder (multiple sclerosis) from other neurologically impaired patients by use of the Luria-Nebraska Battery. Marvel, Golden, Hammmeke, Purisch, and Osmon (1979) examined the effects of age and education upon the Luria subscale indices and determined that both factors significantly affected scores. Subsequently, a correction factor was built into the scoring procedures to adjust for the effects of age and education.

The appeal of the Luria Battery appears to be growing. In Michigan, for example, a study of alcoholic patients is being conducted to determine whether a distinct pattern of scores emerges. In Illinois a comparison of the Luria Memory subscale and the Wechsler Memory Scale is in process. More research with diverse populations will no doubt appear in the literature in the next few years.

In spite of its appeal, the test has been criticized. Reviewers of the test and its approach (Adams, 1980a, 1980b) have pointed out that its scoring and administration methods remain subjective. Adams also has questioned the selection of subjects (their diagnoses and use of psychotropic drugs), and the method of data analysis (use of multiple t-tests) in the validation studies. The test's author (Golden, 1980) has responded that the revised
test manual provides objective scoring and administration criteria. He also provided previously unpublished information on the subject selection process for the validation studies and defended his data analysis procedures as necessary in the first stages of test validation. While admitting that further research is needed, Golden maintained that the methods and results in the validation studies were sound and thus criticisms of the test by its opponents have been unfounded.

Neuropsychological testing and the mentally retarded. There has not been an abundance of research in the application of neuropsychological testing to the understanding of mental retardation. Only two examples of such efforts were found in the literature: a series of studies by Matthews and Reitan using the Halstead-Reitan (1961, 1962, 1963) to compare the mentally retarded with other brain injured groups, and an attempt by Clausen, Lidsky, and Sersen (1976) to differentiate groups of mentally retarded subjects on the basis of diverse measures. The reason for this shortage of research is unclear, since it appears that an understanding of brain-behavior relationships in the mentally handicapped should assist in understanding both the normal and abnormal functioning of the brain. Perhaps one reason is that focus in neuropsychological testing has been maintained upon the diagnosis of the
presence of brain damage. For the mentally handicapped this appears an unnecessary exercise. With input from the Luria school of thought, however, it is hopeful that focus can shift away from the dichotomous diagnosis and toward an analysis of patterns of strengths and weaknesses for groups and individuals.

The research by Matthews and Reitan was designed to explore the usefulness of the Halstead-Reitan test with institutionalized mentally retarded persons. Attempts to normalize the battery for this population were difficult, due to the often generalized and chronic nature of the impairment of the subjects. In the practical vein, many of the Halstead-Reitan subtests were just too difficult for the retarded subjects to comprehend and complete. For example, the Tactual Performance Test, a difficult form-board task which must be completed while the subject is blindfolded, was beyond the ability of many of the subjects (Matthews, 1974). Nevertheless, some results were obtained. Matthews and Reitan (1961) compared mentally retarded subjects' performance with that of persons who were brain damaged, but who had experienced normal childhood development. On the Categories test of the Halstead-Reitan, which is a test of abstract reasoning, the mentally retarded subjects performed poorer. However, when the abstract principle needed to solve the Categories
items remained constant, the mentally handicapped subjects were better able to show improvement and reduce their errors than were the recently brain damaged group. On a formboard task, there was no difference between the two groups on their success in completing the task, but again the mentally retarded subjects showed better ability to improve their performance over trials. These results were interpreted as demonstrating that mentally handicapped subjects may have greater habilitative potential than the recently brain damaged group (Matthews & Reitan, 1962). These same investigators (Matthews & Reitan, 1963) further discovered that mentally retarded persons who had low abstract ability were best able to perform tasks that were dependent upon experiential and verbal ability, and they often had better Verbal IQ than Performance IQ on the WAIS. On the other hand, mentally retarded persons with good abstract ability did better on non-verbal problem solving tasks, such as are found on the Performance section of the WAIS. In reviewing this series of studies Reitan (1966) concluded that neuropsychological examinations of retarded subjects leads to a better understanding of their deficits. Although intellectual skills are generally depressed, meaningful group and intraindividual variability emerges which could lead to individualized rehabilitative pro- gramming.
No research studies have been published investigating Downs syndrome in particular through neuropsychological testing. In the study by Clausen et al. (1976), thirty-five measures of sensory, perceptual, cognitive and motor skills were employed to distinguish four groups of mentally retarded subjects (PKU, Downs syndrome, familial and brain damaged due to mechanical injury at birth). Although some significant results emerged, this research lacked the use of standardized testing procedures. In the Matthews and Reitan series, the investigators combined all mentally handicapped groups together without consideration for their wide range of etiologies and variations in behavior.

With respect to this issue Davison (1974), however, noted the much wider variances in performance of mentally retarded subjects on neuropsychological tasks in the Matthews and Reitan studies than have been found among normal controls. This discovery runs contrary to the general notion that all mentally retarded individuals only represent a limited and rigid range of adaptive abilities, and thus suggests the differentiation of categories within the mentally retarded population:

These data agree . . . that mental retardates are individuals not merely of low adaptive ability in general, but are actually suffering from many different patterns of adaptive insufficiency. Presumably, these patterns have implications both for under-
standing and treatment of this class of individual. It would be instructive to analyze all the protocols, as Matthews has done for the three he presents, to determine how many contain patterns of relatively high and low scores which would support inferences of areas of more or less adequate brain tissue (Davison, 1974, pp. 357-358).

Davison thus implies that the neuropsychological study of the mentally retarded individual is a fruitful endeavor. It is unfortunate that the benefits of such investigations have not been more obvious and provided the impetus for more research.

Summary. The study of brain-behavior relationships at present is far beyond that first proposed by the early localizationist theorists. Present approaches center more around methodological disputes over the use of neuropsychological test batteries than basic theoretical considerations. The new Luria-Nebraska Battery attempts to wed the Luria qualitative approach to the American quantitative approach. The Luria test was also designed to offer time efficiency and simplicity in the administration of neuropsychological tests.

The validation literature suggests that the Luria test can achieve its goals of detecting the presence of brain damage and lateralizing the deficit. Although not a goal which was explicitly stated by the test's authors, the Luria-Nebraska may also provide a tool which is better than the Halstead-Reitan for investigating brain-behavior
relationships in the mentally retarded. If so, the goals of applying a comprehensive theory and related diagnostic method to the study of Downs syndrome is made possible. The final step in this process is to determine whether treatment strategies can be developed from the theory and diagnosis. This is the task of rehabilitation.

The Applications of Human Neuropsychology

Problems in generalizability. The field of neuropsychology appears to hold promise for the future understanding, diagnosis and treatment of brain-behavior relationships in general. However, as the practice of neuropsychology becomes more widespread, clinicians are becoming more aware of the diverse factors which affect results of neuropsychological testing and thus affect the generalizability and practical application of such results.

Sex and age differences have been found in neuropsychological test results. Witelson (1976) found that in normal right handed children between the ages of six and thirteen years, boys had significantly better tactual-spatial scores than girls, and left hand better than right hand performances. Girls had no differences between the two hands. Witelson concluded that the right hemisphere of females may be less specialized for spatial processing than the right hemisphere of males, and thus transfer of function after injury in females may occur more readily.
Age factors are also considered important in analyses of neuropsychological test results. Test performances of normal persons above age fifty often fall into the brain damaged range. Bak and Greene (1980) administered the Halstead-Reitan to two groups of normal adults, aged fifty to sixty-two, and sixty-seven to eighty-six. They discovered that age accounted for 14 percent to 33 percent of the variability between group means on portions of the test, and suggested that new cut-off scores for subjects past age fifty are needed for the Halstead-Reitan.

Parsons and Prigratano (1978) have suggested that in addition to age and sex, level of education, socioeconomic status and the level of sophistication of the examiner are factors to be considered in neuropsychological testing. Conditions related to the brain injury itself may also affect test results. These conditions include duration of time since the insult, age at onset, type of injury, the patient's hand dominance, and his emotional responses to the injury. Thus before any useful applications of neuropsychological testing are derived, it is important to consider these various factors.

For the mentally handicapped the conditions related to the injury may often be controlled by the fact that a majority of retarded individuals have had impaired
brains since birth. The injury is very often chronic and generalized in nature. Sex, age and education, however, should be carefully controlled in neuropsychological studies with the retarded so that generalization implications are clear.

**Neuropsychology and rehabilitation.** The unique quality of neuropsychological testing is the link between diagnosis and rehabilitation (Boll, 1977). Luria emphasized this orientation in his writings and made numerous references to the practical applications of neuropsychology. One example provided (Luria, 1979) is the rehabilitation of a Parkinson's patient. This individual could not walk across a room without a tremor, but was able to do so once pieces of paper were placed on the floor to provide visual cues. Luria proposed this as an example of the flexible reorganization of behavior on a different level once an insult has occurred to one portion of a functional system.

This example is credible if one approaches the task of rehabilitation from a Luria point of view. When insult or disability occurs in one part of the brain, the complex operation of a functional system is disrupted, as are the patient's motor and spatial systems in the example given. Reconstruction of the functional system can, however, occur through substitution of the work of more basic
systems of the brain for the function that has been lost, or by using higher cortical systems to replace the lost function. The integrity of the other portions of the brain which are used to form the new functional system is required. In the example, the patient's intact visual system provides support to the impaired systems so that the task can be accomplished. It is because of this emphasis upon the understanding of the flexibility of the functional systems that Golden (1978) has drawn a distinction between neuropsychological rehabilitation and other forms of rehabilitation. In other forms of rehabilitation the Parkinson's patient would be trained in motor activity without the additional support of the visual cues. Thus training in response to overt symptoms would be emphasized. In contrast, the goal in neuropsychological rehabilitation is the training of basic functions, which can then generalize to the improvement of functioning of several systems as well as restore a lost ability.

There has been little empirical research on neuropsychological rehabilitation. One major reason for this is that the complexity of each person's difficulties makes it impractical to employ the traditional group design of research. Other problems with this type of research include the need to account for numerous patient variables which clinicians in the field are discovering. These factors include sex, age, general health and brain integrity,
social environment of the patient, motivational factors, dominance (since mixed dominance patients show greater recovery potential) and time since the injury. Such a diversity of factors provides uniqueness to each and every case examined. Because of this, some authors have suggested that the single case design, with replications, would be an appropriate approach to rehabilitation research (Golden, 1978; Parsons & Prigratano, 1978).

Nevertheless, Golden (1976, 1978) has maintained that these problems do not inhibit the role of neuropsychologists in the remediation of the neurologically impaired. The neuropsychologist can provide a detailed assessment of the patient's strengths and weaknesses, of his emotional response to the injury, and the interaction of these two aspects. He is able to determine which of the patient's disabilities can be handled initially and most productively so that the patient can have an early experience of success. Another aspect in contributing to the carrying out of rehabilitation plans is the neuropsychologist's ability to stress basic skill training as an avenue to achieving greater generalization.

Rehabilitation of the mentally retarded. Although the field of rehabilitation for those suffering from focal and acute neurological damage appears hopeful, there is generally less optimism for the chronically, generalized
impaired person, such as the mentally retarded. Rehabilitative potential depends upon the integrity of other portions of the brain in order that the other parts can be reorganized to restore the lost function. In all but the most mild forms of mental retardation, it is suspected that large portions of the brain are impaired, and thus there are fewer intact portions which can be utilized.

Yet, even though normality may never be fully achieved in these people, there is no evidence to suggest that their general level of abilities cannot be improved. Special education advances have resulted in improved functioning for many retarded individuals. An example is that of the non-verbal child who, through being taught sign language, has spontaneously begun to verbalize as well. In this case reliance upon the more intact, although still impaired motor system has produced a reorganization capable of producing speech. Knowledge of the generally greater level of intactness of the motor system is a necessary component of developing such a plan for encouraging speech in a non-verbal retarded child.

A second example is the teaching of complex assembly jobs to the mentally retarded adult in a workshop setting. In successful programs of this type it is recognized that the task must be broken down into small, discrete steps, visual cues must be provided, and training
should follow a careful step-by-step process with reinforcement for success. The visual cues and repeated motor activity serve to replace complex verbal instructions and assist the mentally retarded person to learn the complex task. In this instance, an explanation for the known success of a teaching technique is provided through a neuropsychological orientation. The more intact motor and visual systems produce faster learning than does the highly impaired system for comprehending complex language.

The literature on Downs syndrome provides one further example of successful remediation with the retarded. Sidman and Cresson (1973) reported a successful attempt to teach two Downs syndrome persons to read by training a cross-modal transfer using both auditory and visual channels. The subjects were first taught to match printed words to each other, then to match dictated words to pictures, and finally to match dictated to printed words. With the accomplishment of this last step the subjects were able to read the printed word with comprehension of its meaning. The use of these multiple steps may not be necessary for teaching normals to read. For the retarded, however, the additional input is essential.

There is no doubt that there is much more to be explored about the mentally handicapped and the best techniques to result in their improved learning. The final
goal in such explorations is the total improved functioning of the individual. If the Luria approach to neuropsychology and the Luria-Nebraska diagnostic procedure are applied to this end, increased understanding and rehabilitation of a unique group of individuals, such as those with Downs syndrome, may be the end result.

**Summary.** The practical applications of neuropsychology involve the creation of techniques other than those used by non-impaired persons to learn a task. Rehabilitation professionals have been successful in applying these techniques to restore the functioning of patients with focal brain impairment. The task of rehabilitation of the mentally retarded is different, however, due to the generalized nature of their impairment. Yet it is a manageable task, if viewed with the Luria orientation of understanding patterns of strengths and weaknesses and improving present skills rather than attempting to eliminate totally the effects of the handicap.

**Hypotheses**

The concept of adult development, both normal and pathological, is a relatively new area of investigation which became the focus of the present study. The neuropathological findings of degenerative brain tissue in Downs syndrome persons above age thirty-five (Burger & Vogel, 1973; Callner, Dustman, Madsen, Schenkenberg, &
Beck, 1978; Ellis, McCulloch & Conley, 1974; Jervis, 1948; Jervis, 1970; Malamud, 1972; Ohara, 1972; Olson & Shaw, 1969; Solitaire & Lamarche, 1966) suggested that an abnormal developmental aging process is associated with the genetic disorder. Investigators have found it difficult to diagnose the onset of this disorder because medical tests are inadequate for this purpose. Additionally, few theoretical and empirical explorations of brain-behavior relationships in the mentally retarded have been initiated.

It has been suggested that the Luria model of neuropsychology and the resulting Luria-Nebraska Neuropsychological Test would provide the tools for diagnosing the early cerebral aging process in Downs syndrome. This led to the hypothesis that older Downs syndrome persons, whose cerebral aging process was already activated by genetic components, would perform more poorly on the neuropsychological testing than would a young group of Downs syndrome persons. In the present study it was predicted that this effect would be revealed in more impaired scores for the older group on the Luria-Nebraska summary indices.

A shift to the Luria orientation to neuropsychology also suggests a re-orientation in methods of describing and diagnosing disorders such as Downs syndrome. In the Luria model, the focus would become one of understanding
the relationships between brain functioning and specific observed behaviors. There has been an abundance of literature on the measurement of observed behaviors of the mentally retarded, and various behavioral scales have been published for this purpose. However, these measures have never been related to theories of brain impairment, particularly the Luria theories of brain functioning.

In the present study it was predicted that a negative relationship would emerge between more traditional measures of behavior of retarded subjects and neuropsychological measures. High levels of behavioral skills would thus correlate with low levels of brain impairment. This relationship was predicted for comparisons of the Downs syndrome subjects' Luria-Nebraska scores and a measure of overt daily behavior, the Adaptive Behavior Scale published by the American Association on Mental Deficiency (AAMD). It was also predicted that a similar relationship would emerge for the two age groups of Downs syndrome subjects.

Finally, it has been noted that specific brain-behavior patterns have been hypothesized in the literature on Downs syndrome, but these patterns have not been analyzed through neuropsychological testing. Two such patterns are higher Performance IQ than Verbal IQ on the WAIS, and higher receptive language than expressive lan-
guage skills. It was proposed that these patterns would be confirmed by the Luria-Nebraska test results, for the entire group of subjects and for each age group. The Verbal IQ-Performance IQ hypothesis was tested by comparing the Right Hemisphere and Left Hemisphere scales on the Luria-Nebraska and it was predicted that the Left Hemisphere score would be more impaired. The language hypothesis was tested by comparing the Receptive Language and Expressive Language subscale scores on the Luria and it was predicted that the Expressive Language score would be more impaired.

1In all analyses the empirically derived Right Hemisphere and Left Hemisphere scales were used. In the test these are denoted as the R* and L* scales. For convenience, these scales will subsequently be referred to as the Right Hemisphere and Left Hemisphere scales.
METHOD

Subjects

Twenty-five adult female residents of the Mount St. Joseph Home, a residential Intermediate Care Facility for the mentally handicapped in Lake Zurich, Illinois participated in the study. All subjects were diagnosed with Downs syndrome early in their childhood and all were between the ages of twenty and fifty years. All subjects had had similar special educational training and many had received their education at the same residential facility for children. It was important that subjects were not so severely impaired that they would be unable to participate in the experimental procedures. Thus further selection criteria were imposed:

1. The IQ in the mild or moderate ranges of mental retardation, as measured by WAIS Full Scale IQ's. The American Association on Mental Deficiency specifies these ranges at IQ's from 35 to 69 (Grossman, 1973).

2. The absence of severely limiting physically handicapping conditions which would impair the subject's ability to participate in the testing. Such conditions
were blindness, loss of hearing, or loss of use of two or more limbs.

3. The presence of functional language skills, as defined by the subject's ability to carry on a simple and comprehensible conversation with other adults.

The subjects were divided into two age groups in order to do a cross-sectional analysis (Achenbach, 1978). The younger group consisted of twelve subjects between the ages of twenty and thirty-five, and the older group consisted of thirteen subjects between the ages of thirty-six and fifty. Means and standard deviations for WAIS IQ and age for each age group are provided in Table 1. The mean IQ scores for the two groups were not significantly different: \( t(23) = 1.25, p = .11 \) (one-tailed test).

**Materials**

Data were collected through the administration of the Luria-Nebraska Neuropsychological Battery and the AAMD Adaptive Behavior Scale.

The Luria-Nebraska Scale consists of 269 individually administered items which are divided into 14 subscales. The items were derived from the writings of A. R. Luria and the publication of Luria's procedures by Anne-Lise Christensen (1975). The 14 subscales measure different dimensions of performance (Golden et al., 1979).
Table 1
Means and Standard Deviations for Age and WAIS IQ for Each Age Group

<table>
<thead>
<tr>
<th>Group</th>
<th>n</th>
<th>M</th>
<th>SD</th>
<th>M</th>
<th>SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Older</td>
<td>13</td>
<td>40.77</td>
<td>4.44</td>
<td>44.92</td>
<td>3.64</td>
</tr>
<tr>
<td>Younger</td>
<td>12</td>
<td>30.50</td>
<td>3.68</td>
<td>42.75</td>
<td>4.98</td>
</tr>
</tbody>
</table>
The Motor scale involves simple motor movements of the hands, mouth and tongue. Tasks are performed through imitation of the examiner's movements, through verbal instruction, or through kinesthetic and tactile feedback. Correct performance as well as regulation of motor activities are measured. Construction dyspraxia, a disorder in spatial organization, is measured through simple drawing tasks.

The Rhythm scale requires the analysis of tones presented on a cassette recording. The subject is asked to compare, imitate expressively, and reproduce motorically a variety of rhythmic patterns. This test is particularly sensitive to disturbances of attention and concentration.

On the Tactile scale, individuals are asked to locate and describe how they are touched on various parts of their fingers, hands, and arms while they are blindfolded. They are also asked to identify objects by touch alone.

The Visual scale tasks include simple visual identification of objects and pictures of objects, and the use of more complex visual spatial perception. Subjects are asked to identify shaded and overlapping pictures, tell time, recognize directions, analyze three-dimensional pictures, and rotate figures.
The Receptive Speech tasks include the understanding of simple phonemes, words and sentences. The subject is asked to repeat and discriminate sounds, respond to directions and decipher complex sentences.

The Expressive Speech test requires the subject to repeat sounds and words, to read sounds and words, to repeat sentences, to produce spontaneous speech under different conditions, and to manipulate complex grammatical systems.

On the Writing scale, subject's ability to copy written material and to write from dictation are assessed.

On the Reading test, the subject is asked to read letters, sounds, words and sentences.

In the Arithmetic scale, the subject is asked to copy and read numbers, compare numbers, perform simple and complex mathematical operations. This subscale is particularly sensitive to educational deficits.

Short-term and intermediate memory are assessed on the Memory scale. The ability to memorize a list of seven words, remember material presented visually and verbally, and to associate verbal material with pictures are measured.

The Intellectual Processes scale consists of a series of items selected because of their ability to discriminate
brain damaged from non-brain damaged persons. Some items, like picture arrangements, definitions and mathematical problems, are similar to traditional intelligence test items. Less familiar tasks include picture interpretation, understanding of thematic expressions, and concept analysis.

The Pathognomonic scale is derived from the previous eleven scales and consists of items which are highly sensitive to brain damage and which are rarely missed by non-brain damaged persons. This scale is particularly sensitive to the acuteness of a brain injury.

The Right Hemisphere scale is derived empirically through analysis of the results on the first eleven subscales from patients known to have right hemisphere lesions. It is used to determine presence of damage in the right hemisphere.

The Left Hemisphere scale is also an empirically derived subscale from the scores of patients known to have left hemisphere lesions. Comparison of this scale with the Right Hemisphere scale provides useful data regarding the location of damage within the two cerebral hemispheres.

Each item on the Luria-Nebraska is scored on a three-point scale, with zero indicating normal performance, one indicating mild impairment and two indicating performance typical of brain damaged persons. Each subscale is
summed and the scores converted to a T-score profile which displays comparative levels of performances for the individual on all the subscales. The test manual provides specific criteria for scoring each item (Golden et al., 1979).

It is also reported in the test manual (Golden et al., 1979) that reliability measures obtained using odd/ even split half correlational techniques exceeded .90 for a normal population. In a brain injured population, split half reliability measures ranged from .71 for the Motor scale to .90 for the Writing scale. Correlations of the scorings on the full scale by five pairs of examiners yielded a scoring reliability of .97.

The Adaptive Behavior Scale is a behavior rating scale designed to evaluate the adaptive behavior of mentally handicapped, emotionally maladjusted and developmentally disabled persons. The term adaptive behavior refers to "the effectiveness of an individual in coping with the natural and social demands of his or her environment" (Nihira, Foster, Shellhaas, & Leland, 1974, p. 5).

This scale is divided into two parts. Part One consists of items measuring skills in ten areas important to day-to-day living. The ten areas measured are: Independent Functioning (eating, toilet use, cleanliness,
appearance, care of clothing, dressing and undressing, travel): Physical Development (motor and sensory); Economic Activity (money handling, budgeting, shopping); Language Development (expression, comprehension, social language); Numbers and Time; Domestic Activity (cleaning, kitchen activities); Vocational Activity; Self-Direction (initiative, perseverance, leisure time); Responsibility; Socialization.

Part Two consists of items measuring maladaptive behavior. Because such a measure is not considered relevant to the hypotheses being measured in this research, data from Part Two were not utilized.

Authors of the Adaptive Behavior Scale (Nihira, et al., 1974) suggest that it is an instrument that can be useful to identify areas of deficiency for individuals and training programs, to compare an individual's ratings over time or in different situations, and to assist in making decisions regarding research, training and program development.

The Adaptive Behavior Scale is designed to be completed by nonprofessional staff, the most important criterion being that the evaluator know the subject's daily performance well. In the present study the scale was completed by residential care staff at the facility, who
worked in the residential cottages and were very familiar with the subjects.

To score the Adaptive Behavior Scale each section is summed and then the scores are converted to percentile ranks on a Profile Summary Sheet. The percentile ranks were obtained from normative data on approximately four thousand residents of facilities within the United States. The normative sample was further subdivided into ten age groups from age three to age sixty-nine. Subjects were of both sexes and fell within the ranges of mild, moderate, severe or profound mental retardation according to standardized IQ testing data, i.e., IQ's of 69 or less (Grossman, 1973). Inter-rater reliabilities ranging from .71 to .93 on the ten areas of Part One were obtained on a sample of 133 subjects. The scale has been proven successful in discriminating among subjects with various levels of adaptive behavior (Nihira et al., 1974).

Procedure

All subjects were administered the Luria-Nebraska according to the standardized procedures provided in the test manual. Since some flexibility is permitted in the administration of individual items, in keeping with the qualitative spirit of Luria's investigations, minor modifications which were needed for this particular subject population were provided. The examiner abided by the test
authors' admonitions that such administrative modification should not compromise the original intent of each item. In this light, portions of the Intellectual processes subscale which did not contribute to the Pathognomic, Right Hemisphere or Left Hemisphere scales were not administered, as most of these items were too difficult for the population being tested and thus only contributed limited information. It could reasonably be assumed beforehand that since all subjects were unquestionably mentally handicapped, they would have greatest difficulty with this subscale.

Part One of the Adaptive Behavior Scale was completed by facility staff for all subjects, with no modification from the standardized procedures.
RESULTS

The comparisons of the Luria summary indices for the two age groups performed to determine whether the older group was more impaired produced few significant results. The comparisons were made using both corrected and uncorrected $T$-scores. The correction factor described in the test manual was introduced to counteract simultaneously the effects of education and age variables. Since age was the variable being investigated in the initial hypothesis, it was suspected that the correction factor would counteract any differences between the two groups. Thus the uncorrected $T$-scores were analyzed as well.

The data were analyzed by means of the $t$-test (Hays, 1973). None of the comparisons of the corrected $T$-scores were significant at the .05 level and in the predicted direction (older group $T$-scores higher and thus more impaired than younger group $T$-scores). One comparison, Right Hemisphere, was significant at the .10 level (one-tailed test). The corrected $T$-score means, standard deviations and $t$-values for the two groups are presented in Table 2, and the two groups' corrected $T$-score profiles are displayed in Figure 1.
### Table 2
Comparison of Luria Corrected T-Scores for Older and Younger Groups

<table>
<thead>
<tr>
<th>Luria Subscale</th>
<th>Older Group</th>
<th>Younger Group</th>
<th>t</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
<td>M</td>
</tr>
<tr>
<td>Motor</td>
<td>95.19</td>
<td>10.65</td>
<td>90.10</td>
</tr>
<tr>
<td>Rhythm</td>
<td>87.44</td>
<td>7.47</td>
<td>86.94</td>
</tr>
<tr>
<td>Tactile</td>
<td>91.08</td>
<td>11.82</td>
<td>92.50</td>
</tr>
<tr>
<td>Visual</td>
<td>82.50</td>
<td>6.37</td>
<td>81.25</td>
</tr>
<tr>
<td>Receptive Language</td>
<td>90.62</td>
<td>10.99</td>
<td>102.67</td>
</tr>
<tr>
<td>Expressive Language</td>
<td>108.90</td>
<td>13.28</td>
<td>115.59</td>
</tr>
<tr>
<td>Writing</td>
<td>78.31</td>
<td>4.96</td>
<td>78.50</td>
</tr>
<tr>
<td>Reading</td>
<td>78.31</td>
<td>4.68</td>
<td>80.50</td>
</tr>
<tr>
<td>Math</td>
<td>122.31</td>
<td>7.53</td>
<td>122.29</td>
</tr>
<tr>
<td>Memory</td>
<td>87.88</td>
<td>5.39</td>
<td>91.67</td>
</tr>
<tr>
<td>Pathognomonic</td>
<td>94.23</td>
<td>10.60</td>
<td>90.69</td>
</tr>
<tr>
<td>Right Hemisphere</td>
<td>87.01</td>
<td>5.79</td>
<td>83.06</td>
</tr>
<tr>
<td>Left Hemisphere</td>
<td>98.38</td>
<td>4.64</td>
<td>99.26</td>
</tr>
</tbody>
</table>

Although significant, these results were in the wrong direction, i.e., younger group more impaired than older group.

*p < .10 (one-tailed).
Figure 1
Corrected T-Score Profiles for Age Groups

--- Older Group
--- Younger Group
The uncorrected T-score comparisons, however, produced some interesting results. When not corrected for age and education, the scores of the two groups differed in the predicted direction at the .05 level of significance (one-tailed test) on the Right Hemisphere scale, and at the .10 level of significance (one-tailed test) on the Motor and Pathognomonic scales. As suspected, the correction factor had concealed some of the differences between the two groups. The uncorrected T-score means, standard deviations and t-values are presented in Table 3, and the profiles displayed in Figure 2.

Pearson correlations between the AAMD score and Luria scores were computed to determine whether the predicted inverse relationship between the two measures existed. The AAMD total score was obtained by utilizing a mean percentile score derived from the ten AAMD subscales. The means and standard deviations for the AAMD data are presented in Table 4. There were no significant differences between the two groups on any of the AAMD measures.

For the entire group of twenty-five subjects, significant correlations in the predicted direction (one-tailed test) were obtained between the AAMD total score and the following Luria subscales: Motor, Tactile, Expressive Language, Writing, Reading, Math, Pathognomonic and
Table 3
Comparison of Luria Uncorrected T-Scores
for Older and Younger Groups

<table>
<thead>
<tr>
<th>Luria Subscale</th>
<th>Older Group</th>
<th></th>
<th>Younger Group</th>
<th></th>
<th>t</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
<td>M</td>
<td>SD</td>
<td></td>
</tr>
<tr>
<td>Motor</td>
<td>99.04</td>
<td>9.72</td>
<td>91.38</td>
<td>16.27</td>
<td>1.44*</td>
</tr>
<tr>
<td>Rhythm</td>
<td>95.64</td>
<td>7.12</td>
<td>93.61</td>
<td>10.39</td>
<td>.57</td>
</tr>
<tr>
<td>Tactile</td>
<td>87.85</td>
<td>10.82</td>
<td>86.50</td>
<td>10.06</td>
<td>.32</td>
</tr>
<tr>
<td>Visual</td>
<td>81.73</td>
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<td>78.75</td>
<td>6.87</td>
<td>1.23</td>
</tr>
<tr>
<td>Receptive Language</td>
<td>92.31</td>
<td>6.92</td>
<td>100.67</td>
<td>18.99</td>
<td>-1.49</td>
</tr>
<tr>
<td>Expressive Language</td>
<td>112.19</td>
<td>11.76</td>
<td>115.59</td>
<td>17.39</td>
<td>-.58</td>
</tr>
<tr>
<td>Writing</td>
<td>86.31</td>
<td>4.96</td>
<td>86.50</td>
<td>5.54</td>
<td>-.09</td>
</tr>
<tr>
<td>Reading</td>
<td>84.31</td>
<td>4.68</td>
<td>86.50</td>
<td>5.79</td>
<td>-1.04</td>
</tr>
<tr>
<td>Math</td>
<td>132.31</td>
<td>7.53</td>
<td>132.29</td>
<td>10.52</td>
<td>.00</td>
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<tr>
<td>Memory</td>
<td>88.27</td>
<td>4.49</td>
<td>89.17</td>
<td>4.44</td>
<td>-.50</td>
</tr>
<tr>
<td>Pathognomonic</td>
<td>96.09</td>
<td>9.07</td>
<td>89.03</td>
<td>13.46</td>
<td>1.55*</td>
</tr>
<tr>
<td>Right Hemisphere</td>
<td>85.98</td>
<td>6.61</td>
<td>79.72</td>
<td>8.26</td>
<td>2.10**</td>
</tr>
<tr>
<td>Left Hemisphere</td>
<td>98.55</td>
<td>4.43</td>
<td>98.15</td>
<td>6.87</td>
<td>.17</td>
</tr>
</tbody>
</table>

*p < .10 (one-tailed).

**p < .05 (one-tailed).
Figure 2

Uncorrected T-Score Profiles for Age Groups

Older Group

Younger Group
Table 4
Means and Standard Deviations for AAMD Subscale and Total Percentile Scores for Total Group and for Age Groups

<table>
<thead>
<tr>
<th>AAMD Scale</th>
<th>Total Group</th>
<th></th>
<th>Older Group</th>
<th></th>
<th>Younger Group</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
<td>M</td>
<td>SD</td>
<td>M</td>
<td>SD</td>
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<tr>
<td>Independent Functioning</td>
<td>18.68</td>
<td>5.94</td>
<td>19.00</td>
<td>5.83</td>
<td>18.33</td>
<td>6.30</td>
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<tr>
<td>Physical Development</td>
<td>35.36</td>
<td>15.68</td>
<td>32.23</td>
<td>13.89</td>
<td>38.75</td>
<td>17.38</td>
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<td>Economic Activity</td>
<td>31.24</td>
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<td>31.38</td>
<td>12.09</td>
<td>31.08</td>
<td>15.14</td>
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<td>Language Development</td>
<td>40.36</td>
<td>21.87</td>
<td>38.92</td>
<td>18.25</td>
<td>41.92</td>
<td>25.99</td>
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<tr>
<td>Numbers &amp; Time</td>
<td>47.12</td>
<td>16.11</td>
<td>48.15</td>
<td>13.64</td>
<td>46.00</td>
<td>18.99</td>
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<td>Domestic Activity</td>
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<td>13.14</td>
<td>65.54</td>
<td>11.04</td>
<td>65.92</td>
<td>15.60</td>
</tr>
<tr>
<td>Vocational Activity</td>
<td>38.20</td>
<td>10.00</td>
<td>40.38</td>
<td>9.90</td>
<td>35.83</td>
<td>9.99</td>
</tr>
<tr>
<td>Self-Direction</td>
<td>16.56</td>
<td>9.21</td>
<td>16.62</td>
<td>10.01</td>
<td>16.50</td>
<td>8.70</td>
</tr>
<tr>
<td>Responsibility</td>
<td>66.92</td>
<td>22.42</td>
<td>71.08</td>
<td>24.17</td>
<td>62.42</td>
<td>20.43</td>
</tr>
<tr>
<td>Socialization</td>
<td>33.84</td>
<td>19.87</td>
<td>33.08</td>
<td>17.02</td>
<td>34.67</td>
<td>23.33</td>
</tr>
<tr>
<td>Total</td>
<td>39.40</td>
<td>9.05</td>
<td>39.64</td>
<td>8.88</td>
<td>39.15</td>
<td>9.62</td>
</tr>
</tbody>
</table>
Right Hemisphere. The analysis was repeated for each age group to determine whether the same pattern was consistent with younger and older subjects. The patterns obtained in these subanalyses were quite different from that obtained on the total group and different from each other. For the older group only the Luria Motor scale correlated significantly (one-tailed test) with the AAMD total score. For the younger group the following Luria scores correlated significantly and in the predicted direction (one-tailed test) with the AAMD total score: Motor, Visual, Expressive Language, Writing, Reading, Math, Pathognomonic, Right Hemisphere and Left Hemisphere. The data from these analyses are presented in Table 5. There was no significant correlation between the AAMD total score and age for the total group: \( r = -.14, \text{ns} \); for the older group: \( r = -.10, \text{ns} \); or for the younger group: \( r = -.46, \text{ns} \). There were also no significant differences between the two age groups on the AAMD total score: \( t (23) = .13, \text{ns} \).

The different correlational patterns which emerged for the two age groups suggested that the two groups could be discriminated when the Luria and AAMD scores were analyzed simultaneously. To explore this possibility a discriminant function analysis was computed. The thirteen uncorrected Luria T-scores and percentile scores for each of the ten AAMD subscales were entered into this analysis.
Table 5
Pearson Correlations Between AAMD Total Score and Luria Subscale Scores for Total Group and for Age Groups

<table>
<thead>
<tr>
<th>Luria Subscale</th>
<th>AAMD Total Score for Age Groups</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Total</td>
</tr>
<tr>
<td>Motor</td>
<td>-.56**</td>
</tr>
<tr>
<td>Rhythm</td>
<td>-.24</td>
</tr>
<tr>
<td>Tactile</td>
<td>-.44*</td>
</tr>
<tr>
<td>Visual</td>
<td>-.21</td>
</tr>
<tr>
<td>Receptive Language</td>
<td>-.27</td>
</tr>
<tr>
<td>Expressive Language</td>
<td>-.37*</td>
</tr>
<tr>
<td>Writing</td>
<td>-.43*</td>
</tr>
<tr>
<td>Reading</td>
<td>-.36*</td>
</tr>
<tr>
<td>Math</td>
<td>-.45*</td>
</tr>
<tr>
<td>Memory</td>
<td>-.16</td>
</tr>
<tr>
<td>Pathognomononic</td>
<td>-.43*</td>
</tr>
<tr>
<td>Right Hemisphere</td>
<td>-.35*</td>
</tr>
<tr>
<td>Left Hemisphere</td>
<td>-.28</td>
</tr>
</tbody>
</table>

*p < .05.

**p < .01.
The resulting function discriminating between the two age groups had a canonical correlation of .8695. This revealed that the function had a high level of success in separating the two groups. The squared value of the canonical correlation (.756) revealed that 76 percent of the variance in the two groups was accounted for by the discriminant function. Wilks' Lambda, a value denoting the discriminating power of the variables not entered into the analysis, was .244. This indicated that the remaining unentered variables would have contributed little to the discrimination between the two groups. Chi-square analysis revealed that the Lambda value was significant in its ability to discriminate between the two age groups: chi-squared (10) = 25.387, \( p < .01 \). The resulting discriminant function successfully classified 100 percent of the younger group cases and 100 percent of the older group cases. The data in Table 6 reveals the variables entered or removed at each step of the discriminant analysis and the relative contribution of each variable, as measured by the standardized canonical discriminant function coefficients.

These results were interpreted as demonstrating that in order to differentiate between the two age groups of subjects it is necessary to consider both neuropsychological and adaptive behavior measures. However, the success of the discriminant function analysis must be
Table 6

Discriminant Function Analysis of
Luria and AAMD Scores

<table>
<thead>
<tr>
<th>Step</th>
<th>Variable Entered</th>
<th>Removed</th>
<th>Standardized Canonical Discriminant Function</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Right Hemisphere</td>
<td></td>
<td>*</td>
</tr>
<tr>
<td>2</td>
<td>Receptive Language</td>
<td></td>
<td>-1.21444</td>
</tr>
<tr>
<td>3</td>
<td>Pathognomonic</td>
<td></td>
<td>1.41615</td>
</tr>
<tr>
<td>4</td>
<td>Reading</td>
<td></td>
<td>-1.05866</td>
</tr>
<tr>
<td>5</td>
<td>Responsibility</td>
<td></td>
<td>.91810</td>
</tr>
<tr>
<td>6</td>
<td>Memory</td>
<td></td>
<td>-.88435</td>
</tr>
<tr>
<td>7</td>
<td>Motor</td>
<td></td>
<td>1.29206</td>
</tr>
<tr>
<td>8</td>
<td>Economic Activity</td>
<td></td>
<td>.89095</td>
</tr>
<tr>
<td>9</td>
<td>Right Hemisphere</td>
<td></td>
<td>*</td>
</tr>
<tr>
<td>10</td>
<td>Tactile</td>
<td></td>
<td>.51256</td>
</tr>
<tr>
<td>11</td>
<td>Occupation (Domestic)</td>
<td></td>
<td>.53912</td>
</tr>
<tr>
<td>12</td>
<td>Left Hemisphere</td>
<td></td>
<td>-.63913</td>
</tr>
</tbody>
</table>

*The variable Right Hemisphere was entered at step 1, but removed at step 9 because it did not increase the discrimination when combined with variables at steps 2 through 8. A final standardized discriminant function coefficient is not provided when a variable is removed. An explanation for the removal of Right Hemisphere at step 9 may be the overlap of items between Right Hemisphere and the Luria scales at steps 2, 3, 4, 6 and 7. This occurs because the Right Hemisphere scale is derived from the other Luria scales.*
viewed with some caution. This multivariate procedure tends to dichotomize data based upon small differences. Thus cross-validation with other samples is essential before interpretations concerning true differences between the two age groups can be affirmed.

The profiles for the entire group, using both corrected and uncorrected T-scores are displayed in Figure 3. As predicted the comparison of Right Hemisphere and Left Hemisphere Luria subscale scores for all twenty-five subjects yielded significant one-tailed t-test results in the predicted direction. Thus the corrected Left Hemisphere score was higher and more impaired: \( t (24) = -10.45, p < .001 \). Similar results were obtained for the comparison of the uncorrected Left Hemisphere and Right Hemisphere scales: \( t (24) = -11.22, p < .001 \). These results were further supported by the finding that for all twenty-five subjects the WAIS Verbal IQ scores, which generally relate to left hemisphere functions, were lower and thus more impaired than were the WAIS Performance IQ scores, which relate to right hemisphere functions: \( t (24) = -3.53, p < .001 \) (one-tailed test).

Furthermore, these findings were maintained when the uncorrected Right Hemisphere and Left Hemisphere scores were broken down into the two age groups. These results are displayed in Table 7. As predicted, in all instances
Figure 3
Corrected and Uncorrected $T$-Score Profiles
for Total Group
Table 7
Comparison of Uncorrected Left Hemisphere with Right Hemisphere Scale, and Verbal IQ with Performance IQ for Age Groups

<table>
<thead>
<tr>
<th>Variable</th>
<th>Older Group</th>
<th></th>
<th></th>
<th>Younger Group</th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
<td>t</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Left Hemisphere</td>
<td>98.55</td>
<td>4.43</td>
<td>6.73**</td>
<td>98.15</td>
<td>6.87</td>
<td>11.02**</td>
</tr>
<tr>
<td>Right Hemisphere</td>
<td>85.98</td>
<td>6.61</td>
<td></td>
<td>79.72</td>
<td>8.26</td>
<td></td>
</tr>
<tr>
<td>Verbal IQ</td>
<td>45.92</td>
<td>3.43</td>
<td>-2.09*</td>
<td>44.08</td>
<td>4.72</td>
<td>-4.52**</td>
</tr>
<tr>
<td>Performance IQ</td>
<td>50.69</td>
<td>7.03</td>
<td></td>
<td>48.00</td>
<td>5.06</td>
<td></td>
</tr>
</tbody>
</table>

*p < .05 (one-tailed).

**p < .001 (one-tailed).
the Right Hemisphere score was less impaired than the Left Hemisphere score.

The comparison of uncorrected Receptive Language and Expressive Language subscales for all subjects also yielded significant one-tailed $t$-test results: $t(24) = -0.683$, $p < .001$. Comparable results were obtained for the corrected $t$-scores: $t(24) = -6.08$, $p < .001$. In all instances Expressive Language scores were more impaired than Receptive Language scores, as was predicted. These results were also maintained when the uncorrected Receptive Language and Expressive Language scores were broken down into the two age groups, as is evident from Table 8.
### Table 8

Comparison of Uncorrected Receptive Language with Expressive Language Scale for Age Groups

<table>
<thead>
<tr>
<th>Variable</th>
<th>Older Group</th>
<th>Younger Group</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>M</td>
<td>SD</td>
</tr>
<tr>
<td>Receptive Language</td>
<td>92.31</td>
<td>6.92</td>
</tr>
<tr>
<td>Expressive Language</td>
<td>112.20</td>
<td>11.76</td>
</tr>
<tr>
<td>Receptive Language</td>
<td>100.67</td>
<td>18.99</td>
</tr>
<tr>
<td>Expressive Language</td>
<td>115.60</td>
<td>17.39</td>
</tr>
</tbody>
</table>

*p < .0001 (one-tailed).
DISCUSSION

Research Interpretation

It had been predicted that the older Downs syndrome group would fall within that age range in which increased brain deterioration would begin to occur. Thus it would have been expected that the older Downs syndrome group would perform more poorly on the Luria neuropsychological tasks, which are a measure of the intactness of the brain. The t-test analyses of the data only supported this hypothesis to a limited degree.

One reason for this may have been within the test itself. The Luria test authors have devoted most of their validity studies to determining the effectiveness of the test in discriminating such factors as presence or absence of brain damage, acuteness of the lesion and location of the lesion. They have not devoted their studies to the fine-tuned task of discriminating levels of impairment within an already brain damaged group. It may be that the differences in brain functioning between the two age groups, if present, were so subtle as to be undetectable by means of the neuropsychological test alone.

87
The process by which the subjects were selected for the two groups may also have affected the results. Efforts were made to include only those subjects who would be capable of completing much of the test, so as to obtain as much data from the testing as possible. However, in doing this, only those subjects who were less impaired were selected and this may have affected the character of the older group in particular. Older potential subjects who were not selected may thus have been eliminated precisely because of their greater brain impairment manifested in any number of physical and sensory deficits as well as general poor health. On the other hand, those who were selected for the older group may have been the most intact, most healthy overall, and perhaps the ones whose ability to function was unlikely to have begun deteriorating.

A look at the subjects who dropped out of the study while it was in process reveals some evidence to support the presence of a biased drop-out between the two groups. There was a 23 percent drop-out rate from the original total of thirty-two subjects. While this figure may seem high for many subject groups, it is not considered unreasonable for this highly specialized group. From the younger group one subject was dropped because she would not cooperate with the testing, one because she was too limited in language skills to comprehend even the simple Luria test.
instructions, and one because she was having difficulty with her hearing aid which corrected a partial hearing loss. From the older group one subject failed to cooperate, one was dropped because of a diagnosed secondary psychiatric condition, but two were not tested because of hospitalizations or medical reasons. It thus appears that there was a selection bias, by which the more impaired older subjects were eliminated because of medical problems which prohibited their full participation in the study, while younger group drop-outs were caused by other reasons. However, the medical problems in the older group may have correlated with the degenerative process being examined and this may have contributed to the general lack of significance when the two groups' performances were compared.

This problem will not be easy to overcome in future studies, however. The high incidence of medical difficulties, motor impairments and cognitive deficits in Downs syndrome persons will always impede the experimenter's ability to find adequate numbers of subjects. It is suggested that some further attempt to match subjects in the two age groups on a measure of medical condition or general health be made, so that the less healthy older subjects are not automatically eliminated from the research. A longitudinal study with small groups which are available may be one other solution, but this approach
is still vulnerable to selection biases to avoid undue attrition of subjects. A more controlled approach may be that suggested by Achenbach (1978) by which the cross-sectional and longitudinal designs are combined to counteract the biases of sampling, historical and age effects.

On some Luria subscales the results were in the opposite direction to that which was predicted, i.e., the younger group was more impaired than the older group. This occurred consistently for both uncorrected and corrected score comparisons on the Receptive Language, Expressive Language, Writing, Reading and Memory subscales, and there was virtually no difference between the two groups on the Math scale. An explanation for this may be that the younger group may simply have been exposed to a different type of training in areas of language development and academics which affected their results on many of these subscales, since training procedures have changed much in the past two decades.

Another explanation for the more impaired scores in the younger group may be a selection bias which affected that group in particular. It is possible that the younger group contained individuals who were actually less intact than the older group, and who will not survive as have members of the older group. The results on Table 3 reveal that in many instances the standard deviations of
Luria subscale scores were larger for the younger group than they were for the older group. This would suggest that the younger group was more heterogeneous and represented subjects with diverse levels of ability. This diversity may indicate that the younger group included some already highly impaired individuals who will not survive into their fourth or fifth decade of life.

It must not be overlooked, in spite of these possible problems, that one of the Luria subscales, Right Hemisphere, did produce significant differences between the two groups, and that two other subscales, Motor and Pathognomonic, approached significance. Since the right hemisphere is the area of the brain known to be associated with visual-spatial and visual-motor tasks, it may be that brain degeneration is revealed by a deterioration in the subject's ability to complete these types of tasks. If future investigations with more subjects and the medical controls suggested above produce similar results, then it would be possible to hypothesize that the Luria Right Hemisphere scale, and potentially the Motor and Pathognomonic scales, may be a useful diagnostic tool to determine the onset of brain deterioration in Downs syndrome subjects.

The results of the correlational analyses between the AAMD scores and the Luria scores indicated that AAMD
measures may be more useful in predicting decreased brain functioning for Downs syndrome persons at or below the age of thirty-five than for subjects above that age. For the younger group all correlations were in the predicted direction and nine of the thirteen correlations were significant. All the behaviors measured on the AAMD scale were meant to be observable by non-professional staff who work with such subjects. Thus the reports of staff on decreases in the observed day-to-day functioning of a Downs syndrome person at or below age thirty-five may indicate a related brain deterioration. Similar assumptions cannot be made, however, for Downs syndrome persons above age thirty-five.

Better than either the Luria or AAMD measure alone, however, may be the combination of the two in isolating age-related differences in Downs syndrome adults. For this group of subjects the most powerful measures which separated the groups were related to right hemisphere functions, as the Right Hemisphere scale was selected by the discriminant function analysis as the single most discriminating variable. Other discriminating variables were related to such specific skills as language understanding, reading, responsibility, short-term and immediate level memory, motor coordination, money management, tactile sensitivity and domestic occupational skills.
It appears from these initial analyses that it is possible to distinguish age-related differences in the brain functioning and observable behavior of adult Down syndrome persons through use of the Luria and AAMD measures. The most effective initial approach to this task may be to investigate those skills which are related to the functioning of the right hemisphere of the brain, followed by analyses of selected more specific skills.

Confirmation of these promising results by cross-validation is essential, however, before it can be assumed that a method for predicting brain deterioration in Down syndrome has been developed. Besides analyzing similar data from additional numbers of Down syndrome adults, data from non-retarded samples will assist in confirming the accuracy of the present results. In particular, normals of different age levels above age fifty, both with and without diagnosed Alzheimer's disease, are recommended validation samples. Test results from these samples would help to confirm whether changes in the functioning of older Down syndrome persons are similar to changes experienced by non-retarded Alzheimer's patients. In contrast, such studies may also reveal that the changes experienced by the Down syndrome group are unique to them. Consequently the diagnostic methods
developed would be appropriate only for the chronically brain damaged group.

The highly significant finding that the Left Hemisphere Scale was more impaired than the Right Hemisphere Scale supports the notion that lateralization trends can and do exist even in a chronically brain damaged group such as Downs syndrome. Although it has generally been reported that lower functioning mentally handicapped people produce Wechsler Performance IQ's which are higher than Verbal IQ's, efforts have generally not been made to analyze such results for specific sub-categories of mental retardation. The current findings suggest not only support of this trend for the adult Downs syndrome group. They also suggest that the implied relationship between the two hemispheres also exists, with the right hemisphere being less impaired.

These data also provide a neuropsychological explanation for why Downs syndrome persons have been successful with some types of tasks and not with others. While suffering from impairment on both sides of the brain, these persons are less impaired on the side of the brain which is responsible mostly for visual-motor functioning. They are more impaired on the side of the brain which is responsible for language functions and thus less capable of coping with complex language tasks. It would be pre-
dicted from this that learning could occur more success­fully when the material to be learned is presented through non-verbal rather than through verbal channels. Additionally, the type of conceptualization and abstract thought required in assimilating complex verbal material may be harder to learn than are visual-motor tasks. Experience from workshop types of settings, in which visual-motor learning is prominent, appears to support this theory, since Downs syndrome persons often perform well in these settings.

This lateralization finding is also particularly noteworthy because it was found in a female population. Within the non-brain damaged population, females may have less lateralization trends than do males (Witelson, 1976). This trend appears to be counteracted by the addition of the extra chromosome. It would be valuable to repeat the present study with Downs syndrome males to determine whether the same strong lateralization trend exists.

The final finding, that the Luria Receptive Lan­guage score was less impaired than the Luria Expressive Language score, provides support for the notion of Corn­well (1974) that Downs syndrome persons may understand language much better than they can produce it. Evaluators of language abilities may thus be missing part of the Downs syndrome person's language strengths if they con-
centrate only upon expressive skills such as vocabulary and the production of speech sounds. These data suggest that poor verbal expression is not indicative of poor language understanding for Downs syndrome adults. An explanation for this phenomenon may be found in the levels of structuring required for each type of language. To express language, one must first organize concepts before verbalizing. In receiving language, the meaning is already organized by external sources before it is presented. Thus the Downs syndrome person's language deficits may best be characterized by poor language organization capacity.

This relationship between receptive and expressive language may be a characteristic strength/weakness pattern which is typical only of Downs syndrome, and not found in other mentally handicapped groups. If so, it could be a distinguishing diagnosing trait. Comparisons of Downs syndrome patterns with other groups of mentally retarded subjects would either confirm or refute this as a pattern typical only of Downs syndrome.

The dispute regarding the relationship of language and intelligence is certainly not settled by these results. However, the Luria language scores do reveal that the measurement of language levels in Downs syndrome involves analyses of different, and perhaps separate, types of
language functions. Assessments of neither expressive nor receptive language alone is adequate in order to make inferences about cognition. The complexity of language functions revealed in this data would suggest that a direct inverse relationship between language impairment and cognition level is too simplistic a notion.

Clinical Interpretation

A few comments should be made about the clinical interpretation of the Luria test results for the Downs syndrome sample. The profiles presented in Figure 3 are useful in revealing the clinical meaning of the Luria scores.

In the Luria manual (Golden et al., 1979) it is suggested that an average T-score greater than sixty and an elevated Pathognomonic score are indicators of brain damage. Application of this interpretive rule reveals that there is no question that the Downs syndrome subjects display the pattern of abilities which could be diagnosed as brain damaged. The mean elevations of all the individual subscale scores above a T-score of seventy-five is also indicative of a generalized brain impairment rather than a focal lesion, a finding that is not contrary to the general literature on Downs syndrome.
The peak high scores on the profile occur on the Math, Motor, Rhythm, Expressive and Receptive Language subscales. The language scores have already been analyzed as suggestive of a distinctive pattern of language disability in Downs syndrome. If the high Expressive Language score is considered along with the high Motor score, it could be suspected that the subjects' poor motor skills may be contributing to the poor expressive language ability, since expressive speech requires a motor component. The peak on the Rhythm subscale may indicate an auditory attention deficit, but this cannot be affirmed from these limited results.

The extremely high Math score must be interpreted in light of the subjects' limited educational background. The test authors (Golden et al., 1979) have found that even normals can achieve high scores on the Math subtest because the test is sensitive to educational factors. It is interesting to note, however, that the other subscales which are sensitive to educational deficits, Reading and Writing, are among these subjects' lowest and thus least impaired. It is obvious that something more than the lack of educational benefits contributed to the vast differences in these scores.

Another subscale score, Visual, is among the lowest and least impaired. This finding would agree with the
notion that visual modalities are the most effective ways in which Downs syndrome persons can process information.
The difference between the Right Hemisphere and Left Hemisphere Scales reveals that while the brain impairment in Downs syndrome affects all areas of the brain, there is less impairment in the right hemisphere. This is a characteristic of Downs syndrome which has not been previously confirmed in the literature on the disorder.

In general, the clinical interpretation of the Luria results confirms the notion that Downs syndrome persons have a generalized form of brain damage affecting all areas of the brain, but that the right hemisphere is less impaired than the left hemisphere. The most impaired functions are those involving expressive speech and motor abilities, while the least impaired functions involve visual abilities. Consequently, rehabilitation or training methods for these subjects should be designed to maximize the use of the less impaired visual system and right hemisphere functions to improve the functioning of other, more impaired systems.
SUMMARY

To examine age-related functioning in an adult Downs syndrome population, and to provide data regarding lateralization, language skills and adaptive abilities that may differentiate the Downs syndrome group from other adult mentally handicapped groups, the Luria-Nebraska Neuropsychological Battery and the AAMD Adaptive Behavior Scale were administered to twenty-five institutionalized Downs syndrome women between the ages of twenty and fifty.

On the Luria only one of the thirteen summary indices, the Right Hemisphere Scale, successfully discriminated between two age groups of subjects, those above age thirty-five and those below that age. The reason for the low number of significant findings may have been related to a subject selection bias which eliminated the more impaired older subjects. When the Luria results were correlated with the AAMD results, the predicted inverse relationship between neuropsychological functioning and adaptive skills was found, but only for the younger group. The scores for the Downs syndrome subjects above age thirty-five did not reveal this same inverse relation-
ship. When the Luria and AAMD measures were examined together through a discriminant function analysis it was found that the two age groups could be significantly discriminated through a combination of specific measures from the two tests.

These results were interpreted as demonstrating that discrimination of age-related differences can be made based upon the two measures. This successful discrimination may be the first step in detecting brain-behavior changes indicative of brain degeneration in Downs syndrome persons.

The study also produced evidence of lateralized brain damage in the Downs syndrome group and significantly different performance levels with expressive language and receptive language skills, with the expressive language skills being more impaired. These findings may reveal characteristics which differentiate Downs syndrome from other mentally retarded groups. They also provide insight into learning processes and deficit patterns related to brain functioning for Downs syndrome persons.
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The dissertation submitted by Nancy Beard Scholle has been read and approved by the following committee:

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The final copies have been examined by the director of the dissertation and the signature which appears below verifies the fact that any necessary changes have been incorporated and that the dissertation is now given final approval by the Committee with reference to content and form.

The dissertation is therefore accepted in partial fulfillment of the requirements for the degree of Doctor of Philosophy.

Date: 3/24/87

Director's Signature: Alan S. DeWolfe