Speech-language abilities in Rett syndrome.

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SPEECH-LANGUAGE ABILITIES IN RETT SYNDROME

by

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B.S., East Stroudsburg State College, 1980

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**TABLE OF CONTENTS**

<table>
<thead>
<tr>
<th>Section</th>
<th>Page</th>
</tr>
</thead>
<tbody>
<tr>
<td>Introduction</td>
<td>1</td>
</tr>
<tr>
<td>Prevalence</td>
<td>3</td>
</tr>
<tr>
<td>Descriptive Studies</td>
<td>4</td>
</tr>
<tr>
<td>Rationale and Purpose</td>
<td>12</td>
</tr>
<tr>
<td>Method</td>
<td>13</td>
</tr>
<tr>
<td>Subjects</td>
<td>13</td>
</tr>
<tr>
<td>Materials</td>
<td>14</td>
</tr>
<tr>
<td>Results</td>
<td>16</td>
</tr>
<tr>
<td>Discussion</td>
<td>24</td>
</tr>
<tr>
<td>Critique</td>
<td>25</td>
</tr>
<tr>
<td>Appendix</td>
<td></td>
</tr>
<tr>
<td>Bibliography</td>
<td></td>
</tr>
</tbody>
</table>
INTRODUCTION

Rett Syndrome, a disorder only recently recognized and diagnosed (Hagberg, Aicardi, Dias, and Ramos, 1983) results in profound speech-language, oral-motor and pragmatic deficits as well as a number of other severe developmental problems. This disorder is confined to females who are apparently normal until six to twelve months of age. They subsequently develop a loss of motor and cognitive skills, inappropriate social interactions, deceleration of head growth, and severe language deficits. Originally reported by Andreas Rett in 1966, this syndrome became internationally recognized as a separate diagnostic category following publication of Bengt Hagberg’s article in 1983 (Hagberg, et al. 1983). No discriminatory laboratory test has been developed for diagnosing Rett Syndrome. Precise clinical criteria for identification of patients were designed by Hagberg, Goutieres, Hanefeld, Rett and Wilson (1985). These diagnostic criteria include female sex; normal psychomotor development through the first six, often twelve to eighteen months of life, followed by early behavioral, social and psychomotor regression; normal head circumference at birth; development of communication dysfunction and signs of dementia; loss of acquired purposeful hand skills through ages one to four; hand wringing and clapping stereotypes; appearance of gait
apraxia and truncal apraxia/ataxia through ages one to four; and tentative diagnosis until three to five years of age.

Because of the developmental progression and variability of these criteria, Hagberg and Witt-Engerstrom (1986) proposed a four stage clinical pattern and profile of the typical Rett Syndrome patient. Based only on their observation of fifty patients, they recognized this system provided a "rather crude and simplistic frame for a more systematic registration, thought and approach to the complex clinical pattern of Rett Syndrome patients and their changes." (p. 59). The language, developmental, social and medical characteristics of each of Hagberg and Witt-Engerstrom's stages are outlined below:

Stage I begins at birth and normal development is noted until approximately six to eighteen months of age. General slowing in the child's development is then reported. This slowing is characterized by developmental arrest, changed communicability and eye contact, unspecified personality variation, diminished play interest, unspecified-episodic hand waving and deceleration of skull growth.

In Stage II, an obvious loss of acquired abilities is noted. This stage is described as the "rapid destructive stage" involving developmental deterioration, pseudotoxicity (resembling toxic or poisonous reactions), autistic manifestations, severe dementia, loss of hand skill/use,
clumsy apraxic-ataxic mobility, hyperventilation and behavior problems.

Stage III is the "pseudostationary stage", so named because of the perceived leveling off of the child's abilities. The lowest level for hand use and speech ability has been reached. The patient appears mentally retarded although emotional contact is present. Gait apraxia and jerky truncal ataxia are prominent and epileptic symptoms become common.

Stage IV, the "late motor deterioration stage", is characterized by decreasing mobility, increasing lower motor neuron involvement (scoliosis, trophic foot disturbances), growth retardation, cachexia (state of ill health), and a staring unfathomable gaze. Emotional contact improves. Epilepsy is also less severe than in Stage III.

The stages were envisioned as a continuum with the various characteristics described above delineating each stage from the other.

PREVALENCE

In addition to describing the four stages, Hagberg (1985) also reported on the prevalence of Rett Syndrome. In a study in a section of southwest Sweden, including five counties and the city of Gothenburg, a prevalence of .65 per 10,000 girls (1:15,000) and zero for boys was established. Prevalence is not known for the United States. Dr. Budden reported that 350 cases of Rett Syndrome have already
been diagnosed. Identification has been primarily limited to four medical facilities located in different regions of the United States. As information on Rett Syndrome becomes more disseminated, it is assumed more cases will be reported, resulting in a higher prevalence.

DESCRIPTIVE STUDIES

The descriptive studies of Rett Syndrome, while varying in comprehensiveness, are clearly influenced by Hagberg and Witt-Engerstrom's (1986) criteria. The behavioral and medical components of these stages are each discussed below.

PRE_AND_POST_NATAL DEVELOPMENT

Budden (1986) discussed clinical and laboratory data of thirteen females with Rett Syndrome. All the subjects had previously been seen and diagnosed either as autistic, mentally retarded or cerebral palsied. The age range was 2 years 10 months to 17 years 3 months. Ten out of thirteen mothers reported normal healthy pregnancies. Three out of ten reported minimal spotting during the first trimester. Twelve out of thirteen girls were delivered at term; one girl was delivered at thirty-six weeks gestation. All subjects followed the same sequence of normal development, followed by loss of acquired skills. Earlier onset did not correlate with more rapid degeneration.

In addition, Hillig (1985) reported that possible prenatal genetic causes may include a specific reproducible fracture of an X or another chromosome, or an extranuclear
mutation. Hillig recommended uniform collection of all genetically relevant data including family birth history, questionable symptoms prior to the onset of the syndrome, birth sequence, pregnancy information, contraception, parental contact with possible mutagenic agents, tracing of teratogenic influences, background on grandparents and tracing of consanguity.

**NEUROLOGIC DEVELOPMENT**

A characteristic of the diagnostic criteria for Rett Syndrome is normal neurological development during the first six to eighteen months of life, followed by stagnation of developmental acquisition and rapid deterioration of behavior and mental status (Hagberg, et al., 1983).

Naidu (1985) evaluated seventy females with Rett Syndrome, ranging in age from 2 years 5 months to 34 years 5 months of age. Results of this study indicated the development of a "subacute encephalopathy of a very early onset, maximizing in the second year of life with slow recovery and devastating sequelae." (p. 255).

Riederer, Adams, and Haynes (1985) explored the biochemical aspects of Rett Syndrome in nine patients. Results of this testing indicated that ammonia levels tended to be higher in Rett Syndrome subjects than in a normal control group. The difference, however, was not at a significant level. The authors believed that occasional hyperactivity and stress could account for the elevated
levels. No gross changes to amino acids were noted. In conclusion, it was stated, "Rett Syndrome seems to be characterized by a mutation on X leading to brain atrophy and retarded-reduced pigmentation of the substantia nigra and locus coerulus", (p. 358).

Studies focusing on neurological findings included:

Jellinger and Seitelberger (1986), reported on eight autopsies performed between 1966 and 1984, on patients who fulfilled the diagnostic criteria of Rett Syndrome. Gross examination of the brain revealed "considerable diffuse cerebral atrophy with only mild cortical atrophy and mild internal hydrocephalus." (p. 288). Brain weight was significantly lower than that of the age-matched controls. Light microscopy studies revealed very mild-mild diffuse or scattered loss of neurons throughout the cerebral neocortex.

Riederer et al. (1985) performed brain autopsies on three Rett Syndrome females. Results were similar to those reported by Jellinger and Seitelberger (1986). Histological examination revealed normal appearance of all areas of the substantia nigra which ruled out the presence of readily identifiable gross brain dysfunction. "This finding would suggest a more complex structural disturbance in the organization of the neuronal network or that Rett Syndrome is a metabolic disorder" (p. 358).
FEEDING

Budden (1986) described the abnormal chewing associated with tongue thrusting and involuntary undulating tongue movements of her 13 subjects. Abnormal chewing associated with tongue thrusting and involuntary undulating tongue movements were noted in eleven out of thirteen subjects. Feeding problems included difficulty with chewing, swallowing, choking and regurgitation.

BREATHING

Numerous breathing pattern abnormalities (eg. hypernea, apnea) have been reported in the Rett Syndrome literature. Techniques used to investigate these breathing patterns included polygraphic recording obtained while awake and during spontaneous all-night sleep; EEG; electro-oculography; EMG; oral, nasal and thoracic spirography and oxygen saturation. (Lugaresi and Cirignotti, 1982; Cirignotti, Lugaresi and Montagna, 1984; Glaze, Frost, El Hibri, and Percy 1985; Lugaresi, Cirignotti and Montagna, 1985).

During wakefulness Lugaresi et al. (1985) described respiration as rapid and irregular in amplification and rate. Respiration occurred in pseudoperiodic clusters interspersed with apneic episodes which appeared to be diaphragmatic in origin and of variable duration (up to 120 seconds). While asleep, respiration was regular and rhythmic. The authors suggest that "the peculiar breathing
disorder observed in the Rett Syndrome reflects involvement of the behavioral control system of respiration. This disorder may be part of a general impairment of basic vital functioning (gait, language, swallowing, social relatedness). Onset occurs early in the disease's progression and disappearance or improvement is noted with stabilization occurs." (p. 330).

Lugaresi and Cirignotti (1982) also described two patients with a severe breathing disorder during wakefulness. The disorder was characterized by apneic episodes of variable duration which alternated with profound and irregular respiratory movements.

In 1986, Cirignotti et al. studied four subjects with Rett Syndrome. The description of the breathing pattern was similar to those described in the Lugaresi and Cirignotti (1985) study. Results indicated variability of respiratory irregularity occurred both between and within an individual. Heart rate showed no consistent relationship to the breathing pattern. "Such numerous and profound hypoxic episodes could contribute to permanent neurological damage, thus explaining part of the clinical manifestations of Rett patients. Furthermore, these hypoxic episodes could induce possible clinically significant neurochemical changes" (p. 169).

A study by Glaze et al. (1985), using polygraphic electroencephalographic-video characterization, resulted in
similar findings to Lugaresi and Cirignotti. In addition, they reported a decrease (range 7-15%) in REM sleep in six Rett patients as compared to age matched controls (range 16-28%).

All studies presented above indicate the presence of abnormal breathing patterns, either hypernea and/or apnea, as a significant Rett Syndrome characteristic.

LOSS_OF_EMOTIONAL_CONTACT

A characteristic of Rett Syndrome as stated by Hagberg and Witt-Engerstrom (1986) is their loss of emotional contact. Withdrawal is noted through social regression, development of communication dysfunction and signs of dementia. However, a decrease in autistic/withdrawal characteristics is noted in Stages III and IV as compared to Stage II. A previous study by Hagberg et al. (1983) described these characteristics in combination with stereotypic responses to environmental stimuli.

STEREOTYPY

Loss of purposeful hand skills is reported. Stereotypic hand movements, including wringing, clapping and washing, become apparent by one to four years of age (Hagberg and Witt-Engerstrom, 1986).
AUDIOLOGY

Involvement of the cerebral cortex is suggested by Lenn, Olsheo, and Turk (1986) due to the presence of dementia, loss of language and social interaction. Based on this hypothesis, Lenn et al. (1986) examined Rett Syndrome females' auditory processing skills. Using Visual Reinforcement Audiometry, the subjects were trained to discriminate the pitch of two pure tones and discriminate the direction of change. Detection thresholds for these tasks was 20dB above the ambient noise level. Frequency discrimination at 50dB suprathreshold was 5Hz, indicating an excellent ability to differentiate pitches. However, on a sweep discrimination task with sweeps of 100Hz, performance was at the 50 percent level indicating an inability to generalize a learned task. The results could not be attributed to outside factors. This task can normally be successfully performed by five to eight month old infants.

Pelson (1986) assessed the audiologic status of 15 Rett Syndrome females using routine audiologic measures and brainstem techniques as opposed to VRA techniques. His findings revealed a high frequency of otitis media in this population. Sensorineural hearing loss was reported in two subjects, one case being progressive in nature.

FORMES FRUSTES

In addition to the majority of articles reporting descriptors of the classic form of Rett Syndrome, other
articles have described variations noted in some of their patients.

Holm (1985) described two females who fulfilled Hagberg's diagnostic criteria, together with growth deceleration and precocious puberty. One female exhibited breast budding at six years two months, with menarche occurring at nine years nine months. Other authors (Hagberg, et. al. 1983, Naidu 1985, and Budden 1987) did not report incidences of precocious puberty.

Goutieres and Aicardi (1986) reported on the characteristics of seven patients whose characteristics were similar to but did not fulfill the complete diagnostic criteria. Fundamental differences between these patients and classical Rett Syndrome according to the Hagberg et al. (1985) criteria included: (a) no period of normal development, (b) an absence of a period of behavioral and developmental regression, (c) no period of purposeful hand movements, or (d) maintenance of purposeful hand use beyond three years of age. Two subjects also exhibited intense epileptic activity that occurred at the onset of deterioration. The authors stated that these patients can be regarded "as parts of a continuous clinical spectrum extending from typical Rett's and cases of nonprogressive encephalopathies." (p. 194).

Hagberg and Rasmussen (1986) reported an individual case study of a seventeen year old female who met most of
the diagnostic criteria for classical Rett Syndrome. However, she never lost purposeful hand function, nor did she have the hand wringing component of the syndrome. After four years of age, the subject became increasingly communicative and regained some of her previous skills. The authors labeled these variations as "formes frustes".

This literature review together with personal discussion with Dr. Budden, an advisory board member of the International Rett Syndrome Association, indicates that the primary focus of Rett research has been descriptive and/or causal in nature. Personal communication with Budden and Meek (1987) indicated that only one article to date has addressed the therapy needs of this population.

Hanks (1986) stated that while occupational, physical and music therapy do not deter the progression of the disease, they do assist in maintaining or improving function, preventing deformities providing positioning and mobility, and keeping the females in contact with their environment. Budden (unpublished article) advised that the focus for Rett Syndrome should not be "cure" but rather prevention and remediation.

Rationale and Purpose

At present, the focus of Rett Syndrome research has been descriptive and/or causal in scope. Minimal attention has been centered on the services required by these females to maintain optimal skill levels. There is no data
presently available on the benefits of physical, occupational or music therapy on this population. However, the majority of younger females (Stage II) have been receiving these services. A similar situation is occurring in the communication disorders discipline.

The purpose of this study was to provide baseline data on the speech-language and oral-motor skills of Rett Syndrome females. This preliminary data consisting of (a) the description and documentation of language levels prior to and subsequent to the onset of Rett’s and (b) the description and documentation of oral-motor functioning levels subsequent to the onset of Rett’s, will hopefully provide beneficial information for therapists to determine appropriate intervention services (i.e. therapy versus management).

METHODS

SUBJECTS: Eighteen females ranging in age from 3.11 to 19.4 years of age were selected as subjects for this study based on their Rett Syndrome diagnosis. All subjects were independently assigned to one of Hagberg’s four Rett stages by two recognized experts (S. Budden and M. Meek) in the field. Each rater was given a list of subjects and a copy of Hagberg’s criteria. They then placed each female into the appropriate stage based on present skill levels reported. Comparison of rater assignments revealed reliability of 1.0. All subjects were diagnosed and have been followed at
Crippled Children's Division, the Oregon Health Sciences University. It is believed that these girls constitute the entire Rett Syndrome population in Oregon (Budden, 1987; Meek, 1987).

MATERIALS

To obtain a standardized baseline measure of the subjects' receptive and expressive language, the Early Language Milestones Scale (ELM) (Caplan, 1983); the Vineland Adaptive Behavior Scale (Sparrow, Balla and Cichetti, 1984) and the Sequenced Inventory of Communicative Development (SICD) (Hedrick, Prather and Tobin, 1985) were administered.

In addition to these standardized measures, a test of oral-motor functioning was included. This test was developed by Helen Mueller and adapted by Merry Meek (See Appendix).

The ELM was used to establish pre- and post-onset language development levels for the subjects.

The ELM is a screening instrument that measures auditory expressive, auditory receptive and visual skills. Children from birth to three years of age, as well as older developmentally delayed children with functioning below the three year level, can be screened. Test administration takes approximately one to three minutes. There is no formal criteria for passing or failing the ELM for children above three years of age, since normative data on this population was not obtained.
The ELM was adapted to establish pre- and post-onset language development levels for the subjects. Parents were asked to describe their child's skills before deterioration was noted. Their responses were recorded on the ELM score form. An additional score form was used to record each subject's present performance. Present data was obtained by direct testing, observation and/or parent report.

The Vineland Adaptive Behavior Scale Survey Form was used to determine the subjects' personal and social sufficiency. This scale provided a general assessment of post-onset adaptive behavior useful in determining an individual's areas of strength and weakness. Respondents were the subjects' primary caregiver. Use of this form provided norm-referenced information based on a nationally standardized sample of approximately 4800 handicapped and non-handicapped persons aged birth to 18 years, 11 months. Questioning began at the lowest item for each domain (communication, daily living, socialization and motor skills). Ceiling rules (seven consecutive scores of 0) were applied to each domain. A score of two points was awarded for a yes, usual response. One point was given for a sometime or partial response. If the behavior was never noted, zero points were scored. N and DK were reported if there was no opportunity or the caregiver did not know the answer. In order to assure reliability, a direct questioning approach was implemented, rather than a general
question. By using direct questions the examiner could insure standardized questioning.

The SICD provided information on the pre- and post-onset receptive and expressive language skills. Pre-onset skills were assessed by a parent report format usage for all questions. Post-onset skills were assessed by either direct intervention or parent report.

The age range for this measure is birth to 48+ month level. Administration began with the zero month items for both scales. Testing continued until a ceiling was achieved (three consecutive items failed). Normative data was based on a sample of 252 children.

Test results and interpretation was determined according to manual instructions. Age-equivalent scores were obtained for the Vineland and SICD.

RESULTS

The purposes of this paper were to (a) describe and document levels of language prior to and subsequent to the onset of Rett's, and (b) to describe and document levels of oral motor functioning subsequent to the onset of Rett’s.

Specifically, these assessments provided baseline data on the speech-language and oral-motor skills for the subjects at their current stage of Rett Syndrome. This study covers Stages II, III, and IV because, at present, diagnosis cannot be made until loss of acquired skills is observed.
Stage II Subjects:

As reported previously (Hagberg and Witt-Engerstrom, 1986), Stage II females show an obvious loss of acquired skills such as vocabulary and articulation. Autistic manifestations are noted along with severe dementia and breathing problems.

In this study pre- and post-onset results were obtained by averaging the receptive age equivalency scores of the SICD and the age equivalency scores of the ELM auditory receptive and visual skill scales. Age equivalence on these tests were often expressed as age ranges, for example zero to four months. In this event, the averaging was done for both the lower and upper limits of the ranges independently. The SICD expressive and the ELM auditory expressive scores were similarly analyzed. Scores obtained using the Vineland Scale were not computed into the average score because their age equivalency ranges were of intervals considerably larger than those of the ELM and SICD. These ranges therefore were not used in order to avoid a marked and artificially induced elevation of the other scores.

In Stage II with three subjects ranging in age from 3 years 11 months to 4 years 3 months, post-onset results indicated receptive language skills at the eight month level for all Stage II subjects and at the zero to four month level expressively. A score of zero to four months is the lowest obtainable on either measure.
Based on parent report, two of three subjects had been functioning at a higher level expressively pre-onset. Scores ranged from the ten to twenty month level. Receptive scores ranged from ten to twenty-four months. Subject 2’s prior and post receptive language skills remained at the same level. These results are presented in Table 1.

Stage III Subjects:

Stage III is considered by Hagberg and Witt-Engerstrom (1986) as the lowest stage for speech-language ability. However, as emotional contact begins to improve, the females become somewhat more aware of activities in their environment. This awareness influences the development of pre-speech skills, resulting in higher scores on the SICD and ELM than might be expected for Stage III females as characterized by Hagberg and Witt-Engerstrom.

In this study, results indicated receptive and expressive post-onset language skills were within the four to eight month level for seven out of eight subjects. Subject 9’s expressive skills appeared at the zero to four month level.

Based on parent report, subjects in this Stage had previously performed at ages ranging from the six to twenty month level expressively. Receptively, parents reported skills at the eight to eighteen month levels, pre-onset. These results are presented in Table 2.
Stage IV Subjects:

Stage IV is characterized by cachexia and a staring unfathomable gaze. Epilepsy becomes less severe and emotional contact continues to improve.

In this study, results indicated post-onset receptive language skills at the eight month level for two subjects and at the four month level for the two remaining subjects. Expressive skills appeared at the eight month level for all subjects except one. Subject 12's skills ranged from the zero to four month level. Based on parent report, all subjects in this Stage had previously functioned at the eighteen to twenty month level. These results are represented in Table 3.

Oral-Motor Functioning:

The second goal of this paper pertained to the oral-motor skills assessed post-onset of Rett Syndrome. Assessment involved notation of postural alignment, presence or absence of abnormal patterns and reflexes, dental development and respiratory pattern. In addition, digital stimulation of the facial region was performed. The evaluation was supervised by a recognized specialist in the field. This individual trained a second examiner prior to participation in this field. Reliability was established on classification of severity (mild, moderate, severe) and tonicity (hyper versus hypo). Each examiner would independently assess a child, record their severity and tone.
findings. A comparison of results followed. If there was discrepancy, a joint examination would be completed resulting in a mutual diagnosis. Following training on five subjects, an overall inter-rater reliability of .85 was obtained.

Tables 4, 5 and 6 represent the present postural tone and respiratory skills of the subjects across Stages. In normal development, it takes until approximately eight years of age for children to use a mature abdominal/thoracic respiratory pattern. This pattern requires postural control with a normal spine and normal postural tone through the head and trunk musculature.

The abdominal pattern is used by all infants until the movement component of rotation is established by eight months of age. Use of an abdominal pattern by these subjects is a natural result of abnormal postural tone and movement. The increase of scoliosis or curvature of the spine restricts movement of the rib cage during phases of inhalation/exhalation.

None of the subjects reached the "mature" respiration stage prior to the onset of Rett Syndrome. Stage II subjects 2 and 3 are ambulatory, but both exhibit mild hypotonia and prolonged apneic episodes. Subject 1 is moderately hypotonic and is non-ambulatory. All three subjects use an abdominal respiratory pattern with episodes of hyperventilation.
In Stage III of Rett Syndrome, all subjects except 4 and 8 are mildly-moderately hypotonic. Subject 4 is mildly hypertonic while Subject 8 continues to have normal tone. An immature abdominal-thoracic respiratory pattern is used by all subjects except 6 and 11. Their pattern is abdominal. Hyperventilation and/or apneic episodes are noted in all subjects.

The variation in postural tone becomes severe in Stage IV. Subjects 12 and 15 are hypertonic while Subjects 13 and 14 are hypotonic. Subjects 13, 14, and 15 use an abdominal respiratory pattern with increased spasticity. Clinically the tone through the thoracic region restricts the use of abdominal/thoracic components in respiration. Subject 12 has a shallow immature abdominal/thoracic pattern. This subject has scoliosis rods in her back to provide postural stability. Hyperventilation and/or apneic episodes are noted in all subjects.

Table 5 represents the variations in tone noted in specific oral structures. The presence or absence of the associated characteristics, bruxism and drooling, is also reported according to stage.

In Stage II, all subjects exhibit hypotonicity of postural tone and the tongue. Subjects 1 and 2 also are hypotonic in the cheeks. Subjects 1 and 2 are presently drooling and grinding their teeth.
All subjects in Stage III exhibit hypotonicity of their postural tone and the tongue, except Subject 4 whose postural tone is hypertonic and Subject 8 whose tone is normal. Subjects 5, 8, 9, and 10 cheeks are also hypotonic. Subject 7 exhibits hypotonicity of both the lips and cheeks. Bruxism is noted on Subjects 7 and 10. Drooling is noted in all subjects except 4 and 10.

Considerable variation in tonicity in Stage IV was noted. Hypo- and hypertonicity in the oral structures was found. Subjects 12, 13 and 14 exhibit hypertonicity of the tongue. Vasculations are also noted in these subjects. Hypertonicity of the lips and cheeks is present in Subjects 12 and 14. Subject 15, however, exhibits hypotonicity of tongue but hypertonicity of the cheeks. No instances of bruxism or drooling are noted in Stage IV subjects.

Table 6 presents the feeding abilities of these subjects. No pattern of skills according to stage is noted. Six of the fifteen subjects are maintained on a pureed food diet; six have a table food diet; one has a combination of pureed food and gastrostomy tube diet; and one is totally dependent on gastrostomy feedings. The two subjects with gastrostomies appear to be progressing through the stages of Rett Syndrome rapidly and their loss of oral-motor skills is most severe.

Lip closure while drinking is present for the majority of subjects although evidence of tongue protrusion and poor
or unusual movements are also reported. The normal development of the rotary component of chewing is generally observed after eight months of age when the rotational component in postural movement is established. It takes until after three years of age for the refinement of the rotary component to be established for chewing nuts, raw carrots, celery, tough meats, etc. None of the subjects had reached the rotary stage of chewing prior to the onset of Rett Syndrome due to its maturational component. Chewing is usually vertical. Subjects 3 and 4 are the only ones who exhibit rotary chewing at all. Deviation of the tongue to the left is also noted in some subjects. Subject 7 dislocates her jaw to the left while eating.

Table 6 also shows the vocalizations produced with the documented oral skills for lips, tongue and jaw. These productions range from consonant-vowel combinations and vowels to laughter, crying and yelling. The patterns of vocalization are related to the severity of their current oral-motor skills. In Stages II and III, consonants and vowels are produced along with differential sounds (laugh, cries, raspberries, etc.) In Stage IV, only a limited range of vowels is noted.

According to this data, minimal variation in receptive and expressive skills across and within the stages is noted. The range of post-onset performance was from zero to eight months. However, clearly noticeable variation was noted in
Rett Syndrome females' oral-motor skills as indicated by the range of feeding and vocalization abilities reported.

**DISCUSSION**

As stated previously, the purposes of this initial study are (a) to describe and document levels of language prior to and subsequent to the onset of Rett's and (b) to describe and document levels of oral-motor functioning subsequent to the onset of Rett's.

Based on the data obtained using the protocol described in the Methods section, it appears that the post-onset speech-language skills of this population range from less than four to eight months receptively and expressively. Pre-onset skills ranged from eight to twenty-four months. The major decline in acquired skills is noted in Stage II (Hagberg and Witt-Engerstrom, 1986). Therefore, to prevent or lessen this decline, the focus of intervention for a female in Stage II should be maintenance of as much previously acquired skill as possible. By providing special services (i.e. physical therapy, occupational therapy, speech-language therapy, music therapy), perhaps skill levels would not diminish to the pre-symbolic level.

In Stage III, when an increase in emotional contact is noted, intervention should focus on expanding pre-speech skills that capitalize on the regression of autistic-like symptoms. Those skills might include sustained eye contact,
visual tracking, and/or choice selection by focused eye
gaze.

The gains achieved in Stage III should be maintained
for as long as possible in Stage IV. The addition of new
skills is unlikely due to the motor deterioration and
cachexia involved in the final stage. It may become
physically impossible for the individual to carry out new or
even previously performed activities.

While the recommendation of this paper is to provide
routine direct therapy services to this population,
additional research needs to be obtained to substantiate the
gains made in relation to frequency and focus of
intervention. If the females' skills have appeared to
plateau, then appropriate services might include monitoring
and consultation with the classroom teacher as opposed to
direct intervention.

All subjects in Stage II are receiving special
services. A longitudinal study comparing their skills to
the older females who did not receive the same services
could provide information on the benefits of direct
intervention versus monitoring/consultation. This
differential provision of services may relate to the fact
that many of the older Rett Syndrome subjects were initially
diagnosed as severely-profoundly mentally retarded and/or
autistic. As a result they may not have received all of the
services provided to the more recently diagnosed subjects.
In addition to the loss of communicative abilities, these females experience difficulty maintaining nutritional needs due to poor oral-motor functioning. Variations in tone ranging from noticeable hypotonicity/hypertonicity throughout the body, to the presence of both hypo- and hypertonicity within the same body has been noted across stages. Further research needs to focus on a possible recognizable pattern of decline or the development of compensatory patterns (i.e. hypotonic cheeks with hypertonic tongue) based on initial Stage I or II postural tone.

Another research area would be to compare the vasculations and leftward deviations of older and/or Stage IV females with those noted in the geriatric population (Meek, personal communication, January 1987). This data would support or negate the possibility of a rapid aging process associated with Rett Syndrome.

Research should also focus on intervention techniques that might allow these females to maintain a normal diet for a longer period of time. While these females appear to maintain lip closure through Stage III, many never develop rotary chewing and have difficulty controlling the bolus of food with their tongue. Nutritional supplements that may be developed to prevent malnutrition should also be pursued.

Another avenue for research on this study would be to collect multiple data points while the subject is in the same stage. Documentation on the stability or diversity of
skills over time would be helpful in developing goals and maintenance plans.

As stated previously, Rett Syndrome is a recently recognized and diagnosed disorder. The focus of the majority of research and articles has been descriptive or causal in nature. Strengths of this article include (a) the fact that a previously unexplored aspect of Rett Syndrome has been examined and documented. Speech-language levels and pre and post-onset were presented. In addition, post-onset oral-motor skills were described; (b) standardized instruments were administered to the subjects allowing for reduplication of the study; (c) a comparatively large N was obtained considering the low prevalence of this syndrome; and (d) recognized leaders in the fields of Rett Syndrome and speech-language disorders acted as consultants and provided supervision during protocol administration.

Weaknesses in this study include the lack of standardized tests assessing early developmental levels and presymbolic skills (zero to six month level). While minimal post-onset variation between subjects was reported, poor test sensitivity may be a significant factor. With a more precise evaluation instrument, individual differences may become apparent. Also, the addition of informal assessments focusing on home programs and motivational materials used in conjunction with standardized assessments may indicate
individual strengths and weaknesses (i.e. auditory versus visual modality).

Another criticism of this study is the sparse documentation of pre-onset speech-language and oral-motor skills. The use of parent reporting as the sole source of previous skills does not provide an adequate base for determining pre-onset skills. Rater reliability was not determined and variables that may have negatively affected reporting include: lack of awareness of developmental milestones, confusion with other siblings' skills, objectivity, length of time female has been post-onset, and lack of written material to refer to. Substantiation by medical records obtained during well baby care for speech-language skills and tonicity would have allowed for validation of reported pre-onset skills.

In summary, this study provides basic information on a group of Rett Syndrome subjects in the various stages of the disease. Development of appropriate intervention services needs to be a prime focus for individuals involved with this population. Without the implementation of an appropriate management plan for these females, the prognosis for acquiring functional speech-language and oral-motor skills is poor.
APPENDIX

Speech Evaluation

Table 1  Prior and Present Language Ages in Stage II
Table 2  Prior and Present Language Ages in Stage III
Table 3  Prior and Present Language Ages in Stage IV
Table 4  Present Postural Tone and Respiratory Skills
Table 5  Tone Levels in Specific Oral Structures
Table 6  Present Feeding Abilities
Table 7  Raw Data on all Subjects

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SPEECH EVALUATION
Merry M. Meek

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1. Postural tone and alignment

2. Changes in facial expression

3. Response to finger stimulation
   Outside mouth
   Inside mouth

4. Dental development
   Bite
   Teeth
   Gums

5. Oral reflexes
   Rooting reflex
   Suck/swallow reflex
   Bite reflex
   Gag reflex

6. Feeding behavior
   Coordination of suck, swallow, breathing
   Normal biting
   Munching-vertical chewing
   Rotary chewing
   Lip closure during swallow
   Jaw grading
   Drooling

7. Abnormal patterns
   Abnormal bite (tonic)
   Jaw thrust
   Jaw extension
   Tongue thrust
   Tongue protrusion

8. Respiration pattern
   Abdominal
   Immature abdominal-thoracic
   Mature abdominal-thoracic
   Asynchronous

9. Results of videofluoroscopy
10. Tongue
   At rest
   In Motion

11. Respiration
    At rest
    In motion

12. Voice
    Pitch, range
    Loudness
    Rate: Intonation
Table 1: Prior and Present Language Age in Stage II of Rett Syndrome

<table>
<thead>
<tr>
<th>Subject 1</th>
<th>Subject 2</th>
<th>Subject 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>CA 4yr 3mo</td>
<td>CA 4yr 3mo</td>
<td>CA 3yr 11mo</td>
</tr>
</tbody>
</table>

- ◦ Prior Receptive
- ▲ Prior Expressive
- ○ Present Receptive
- △ Present Expressive

N.B. The symbols are placed at the ceiling level for each subject. Common ranges for test scores are 0-4 months and 4-8 months.
Table 2: Prior and Present Language Age in Stage III of Rett Syndrome

Prior Receptive  Present Receptive
Prior Expressive  Present Expressive

N.B. The symbols are placed at the ceiling level for each subject. Common ranges for test scores are 0-4 months and 4-8 months.
### Table 3: Prior and Present Language Age in Stage IV of Rett Syndrome

<table>
<thead>
<tr>
<th>Subject</th>
<th>CA</th>
<th>Prior Receptive</th>
<th>Present Receptive</th>
<th>Prior Expressive</th>
<th>Present Expressive</th>
</tr>
</thead>
<tbody>
<tr>
<td>12</td>
<td>3mo</td>
<td>🔄</td>
<td>○</td>
<td>🔄</td>
<td>○</td>
</tr>
<tr>
<td>13</td>
<td>3mo</td>
<td>○</td>
<td>🔄</td>
<td>○</td>
<td>🔄</td>
</tr>
<tr>
<td>14</td>
<td>11mo</td>
<td>○</td>
<td>Δ</td>
<td>○</td>
<td>Δ</td>
</tr>
<tr>
<td>15</td>
<td>11mo</td>
<td>○</td>
<td>Δ</td>
<td>○</td>
<td>Δ</td>
</tr>
</tbody>
</table>

- 🔄: Prior Receptive
- ○: Present Receptive
- Δ: Prior Expressive
- ○: Present Expressive

The symbols are placed at the ceiling level for each subject. Common ranges for test scores are 0-4 months and 4-8 months.
Table 4: Present Postural Tones and Respiratory Skills For All Subjects

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<tr>
<th>Postural Tone</th>
<th>Respiration</th>
<th>Hyperventilation</th>
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<tr>
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<td>moderate hypotonia</td>
<td>abdominal</td>
</tr>
<tr>
<td>2</td>
<td>mild hypotonia</td>
<td>abdominal</td>
</tr>
<tr>
<td>3</td>
<td>mild hypotonia</td>
<td>abdominal</td>
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<tr>
<td><strong>Stage III</strong></td>
<td></td>
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</tr>
<tr>
<td>4</td>
<td>normal to mild hypertonia</td>
<td>immature abdom.-</td>
</tr>
<tr>
<td>5</td>
<td>moderate hypotonia</td>
<td>immature abdom.-</td>
</tr>
<tr>
<td>6</td>
<td>mild hypotonia</td>
<td>abdominal</td>
</tr>
<tr>
<td>7</td>
<td>mild hypotonia</td>
<td>immature abdom.-</td>
</tr>
<tr>
<td>8</td>
<td>normal</td>
<td>immature abdom.-</td>
</tr>
<tr>
<td>9</td>
<td>mild hypotonia</td>
<td>immature abdom.-</td>
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<tr>
<td>10</td>
<td>mild hypotonia</td>
<td>immature abdom.-</td>
</tr>
<tr>
<td>11</td>
<td>normal to mild hypotonia</td>
<td>abdominal</td>
</tr>
<tr>
<td><strong>Stage IV</strong></td>
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<td></td>
</tr>
<tr>
<td>12</td>
<td>severe hypertonia</td>
<td>shallow immature</td>
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<tr>
<td>13</td>
<td>severe hypotonia</td>
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<tr>
<td>14</td>
<td>severe hypotonia</td>
<td>abdominal</td>
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<tr>
<td>15</td>
<td>severe hypertonia</td>
<td>abdominal</td>
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Table 5: Tone Levels in Specific Oral Structures With Associated Behaviors

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<tr>
<th>Stage</th>
<th>Postural Tone</th>
<th>Tongue</th>
<th>Lips</th>
<th>Cheeks</th>
<th>Bruxism</th>
<th>Drooling</th>
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<tr>
<td>2</td>
<td>↓</td>
<td>↓ deviates left</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>↓</td>
<td>↓</td>
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<td></td>
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<td></td>
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<tr>
<td>4</td>
<td>normal to ↑</td>
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<td></td>
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<td>5</td>
<td>↓</td>
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<tr>
<td>6</td>
<td>↓</td>
<td>↓ deviates left</td>
<td></td>
<td></td>
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<tr>
<td>7</td>
<td>↓</td>
<td>↓</td>
<td></td>
<td></td>
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<tr>
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<td>↓</td>
<td>↓</td>
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<tr>
<td>11</td>
<td>normal to ↓</td>
<td>↓</td>
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<tr>
<td>IV</td>
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</tr>
<tr>
<td>12</td>
<td>severe ↑</td>
<td>↑ vasculations</td>
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<td>↑</td>
<td>↑</td>
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<tr>
<td>13</td>
<td>severe ↓</td>
<td>vasculations↑</td>
<td>↓ deviates left</td>
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</tr>
<tr>
<td>14</td>
<td>severe ↑</td>
<td>↑ vasculations↑</td>
<td></td>
<td>↑</td>
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<tr>
<td>15</td>
<td>severe ↑</td>
<td>↓</td>
<td></td>
<td></td>
<td>↑</td>
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</tbody>
</table>

**Code**

- ↑ = hypertonia
- ↓ = hypotonia
- ✓ = presence

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<table>
<thead>
<tr>
<th>Subject</th>
<th>Feeding Textures</th>
<th>Lip</th>
<th>Tongue</th>
<th>Jaw</th>
<th>Reflexive Sounds</th>
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<td>no</td>
<td>poor labial mvt.</td>
<td>munch/ neutral</td>
<td>loud guttural sounds</td>
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<td>closure</td>
<td>suckle and protrusion</td>
<td>munch</td>
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<td>table food bottle fed liquids</td>
<td></td>
<td></td>
<td>rotary</td>
<td>some CV + gestures</td>
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<tr>
<td>III 4</td>
<td>table foods</td>
<td>closure</td>
<td></td>
<td>vertical</td>
<td>laughs, cries</td>
</tr>
<tr>
<td>III 5</td>
<td>pureed foods</td>
<td>closure</td>
<td>protrusion</td>
<td>vertical</td>
<td>giggles</td>
</tr>
<tr>
<td>III 6</td>
<td>pureed foods</td>
<td>closure</td>
<td></td>
<td>munch/ vertical</td>
<td>h p +vowel sounds</td>
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<tr>
<td>III 7</td>
<td>pureed + gastrotomy</td>
<td>closure</td>
<td></td>
<td>dislocates to left</td>
<td></td>
</tr>
<tr>
<td>III 8</td>
<td>likes crunchy food</td>
<td>closure</td>
<td></td>
<td>vertical extension</td>
<td>yells, cries</td>
</tr>
<tr>
<td>III 9</td>
<td>table foods</td>
<td>closure</td>
<td>protrusion</td>
<td>vertical</td>
<td>noises &amp; gestures</td>
</tr>
<tr>
<td>III 10</td>
<td>table finger food</td>
<td>closure</td>
<td>protrusion when drinking</td>
<td>vertical</td>
<td>lala, tongue play, laughs</td>
</tr>
<tr>
<td>III 11</td>
<td>table food chokes on liquids</td>
<td>closure</td>
<td>exaggerated suckle, tongue lateral</td>
<td>vertical</td>
<td>signs</td>
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<td>Lip</td>
<td>Tongue</td>
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<tr>
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<td>pureed</td>
<td>poor closure</td>
<td>deviates left weak bite</td>
<td>high pitched vowels, staccato type sounds</td>
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<tr>
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<td>pureed</td>
<td>poor closure</td>
<td>deviates left</td>
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<tr>
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<td>gastrostomy aspirates liquids</td>
<td></td>
<td></td>
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<td></td>
</tr>
<tr>
<td>15</td>
<td>table foods</td>
<td>protrusion with semi-solids can lateralize</td>
<td>munch/vertical weak bite</td>
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Table 7: Raw Data for Subjects

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<th>Stage I</th>
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<th>ELM</th>
<th>AR</th>
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<th>VINELAND COMM</th>
<th>Soc</th>
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AE -auditory expressive  AR- auditory receptive  V- visual
Comm-Communication  Soc- Socialization

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BIBLIOGRAPHY


