Developmental Guidelines for Children with Myelodysplasia

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A new inventory of 166 milestones and developmental tasks was designed and given to parents of 173 children with myelodysplasia to use over a 2½-year period to record their children’s development. The inventory included items in the categories of self-help (including independent toileting), personal-social development, and gross motor development. The children were placed in one of four subgroups according to their level of paralysis. For each motor-level group, the percentage of those performing each item at various ages was calculated. The ages of routine performance of selected self-help and personal-social items are reported for 20, 50, and 80 percent achievement for the children for whom the data were known. There was a tendency for the less paralyzed children to learn skills more quickly than their more paralyzed peers. However, the wide age ranges within groups for learning individual skills indicate that factors other than the level of paralysis affect the rate of development, and these factors need to be identified. The data provided represent a reasonable estimate of when children with varying degrees of paralysis can first be expected to perform developmental tasks. The tables can be used to establish learning objectives and programs so that more children with myelodysplasia can learn independence at an earlier age.

Key Words: Child development, Myelodysplasia.

Many children born with myelodysplasia now survive to adulthood. The majority have normal intelligence but must learn to adapt to visual perception and hand function impairment, urine and stool incontinence, and reduced mobility. The severity of these handicaps is related to the level of paralysis.

As life-threatening complications have become more easily managed, a major concern for these individuals is that many are growing up inadequately prepared for independent life. Recently, efforts have been directed toward anticipating problems commonly associated with poor personal and social adaptation and toward facilitating normal development. These attempts are hampered by the lack of information that defines how children with myelodysplasia develop. For obvious reasons, norms based on the performance of children without handicaps are inadequate for children with so many special problems that potentially can affect developmental outcome.

Anticipatory guidance is dependent to a large extent on a systematic method of developmental assessment and on valid documentation of change. For purposes of habilitation, a functional profile must be determined that outlines the child’s strengths, weaknesses, and next higher level of development. Ideally, this functional profile should focus on the needs particular to a child’s condition. Systematic functional or developmental assessments have been published for children with cerebral palsy, mental retardation, and visual impairment.
A study of the development of children with myelodysplasia was reported in 1976. The rate of the children’s development was related to their level of motor paralysis, but there was considerable variability of achievement among similarly disabled peers. The developmental curves published have been useful in obtaining a gross assessment of a given child and in counseling parents, who appreciated seeing graphically how their child was functioning. However, there were too few items to adequately cover important aspects of development, and it was not possible with these mean curves to define the earliest age of achievement of many important developmental tasks. In addition, the data did not differentiate between what the children were capable of doing and what they routinely did. This differentiation is important because independent and routine behavior lead to functional independence.

This article will describe a new study defining the earliest age and range of age at which various milestones and developmental tasks are achieved for one sample of children with myelodysplasia. Data presented here will define general self-care and personal-social development.

**METHOD**

**Inventory**

A new inventory based both on clinical experience with children who have myelodysplasia and on preexisting developmental assessments for nonhandicapped children was designed to be completed by parents. It consisted of 166 items listed in developmental sequence and grouped into the following categories: personal-social, eating, dressing, grooming, toileting, gross motor activities, and locomotion. Both simple milestones and more complex sequential tasks leading to functional independence were included in the inventory. (The inventory and predetermined criteria for passing each item are available from the authors.)

One hundred twenty-six of the 166 items required two responses. One response was the age at which the child first was able to perform the item, and the second was the age at which the child performed the item routinely. The remaining 40 items were only applicable for either a first-time or a routine response. In general, a first-time response simply required that the child was capable of performing a task and had been observed to do so. Criteria for a routine response were more strict: the child had to perform the item with no exceptions at all appropriate occasions. The inventory was pretested on 20 parents of children with myelodysplasia, and the parents’ suggestions were incorporated into the final form.

**Subjects**

Eligible subjects were selected by screening a computerized clinical data file of children enrolled at the University of Washington Congenital Defects Clinic. The clinic serves approximately 275 children with myelodysplasia representing the majority of cases in Washington, Alaska, Montana, and northern Idaho. Patients are examined at six-month intervals from birth to two years of age and yearly thereafter to age 21. The type and degree of the child’s involvement in a physical therapy program varies depending on each individual child’s needs and the availability of local resources. The children span the socioeconomic and ethnic spectrums. One hundred seventy-three children were included in this study based on the following criteria: IQ of 70 or above, age between three months and 19 years, controlled hydrocephalus, no spasticity, and no major unrelated disabilities.

Intelligence quotients for children over age three years were determined using the Wechsler Intelligence Scale for Children (WISC) or the Stanford-Binet test. Children three years of age or under were included in the study if they were in the normal range, with the exception of gross motor function, according to the Gesell Developmental Schedule, the Denver Developmental Screening Test, or the Bayley Scales of Infant Development.

Education for each school-age child was categorized as follows: 1) no education or training, 2) private school for handicapped or public school with special placement, 3) normal (nonhandicapped) school placement and more than two academic years delayed for age, and 4) normal school placement and less than two academic years delayed.

Of the 200 parents or guardians who received a questionnaire, 7 declined to participate, 11 were lost to follow-up because they moved out of the area, and 9 were dropped because of unreliable record-keeping or language limitations that prevented complete explanation of criteria necessary for passing the items. Eighty-eight percent of the families were white, 2 percent were black, 4 percent were Native American, 2 percent were Asian, and 4 percent were Hispanic. Sixteen of the 173 parents were foster or adoptive parents. Record-keeping was done by mothers, with the exception of two fathers and two houseparents of boys living in group homes.

Subjects were placed in one of four groups based on the lowest functioning neuromuscular unit determined by the results of a recent manual muscle strength examination (Tab. 1). Table 2 describes the sex ratio, mean IQ, and mean age and range for the children in the four motor-level groups. A one-way analysis of variance showed no significant difference between the motor-level groups and intelligence quotient ($F = 1.51; df = 3, 95; p = NS$). Nor were there
TABLE 1
Criteria for Determining Motor-Level Groups Using Lowest Innervated Muscle Group(s)
(Strengths Approximate Standard Manual Muscle Testing Criteria)

<table>
<thead>
<tr>
<th>Group 1</th>
<th>Group 2</th>
<th>Group 3</th>
<th>Group 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thoracic and Lumbar (L2 and above)</td>
<td>Mid Lumbar (L3, L2-4)</td>
<td>Low Lumbar (L4, L5)</td>
<td>Sacral (S1 and below)</td>
</tr>
<tr>
<td>erector spinæs, abdominals present</td>
<td>good knee extensors, zero knee flexors</td>
<td>good to normal ankle dorsiflexors, medial knee flexors</td>
<td>good hip abductors and extensors, poor to fair plantar flexors</td>
</tr>
<tr>
<td>pelvic girdle elevators present</td>
<td>poor knee extensors, poor knee flexors</td>
<td>good to fair lateral knee flexors, hip abductors and extensors, foot everters</td>
<td>no motor loss, possible incontinence</td>
</tr>
<tr>
<td>hip flexors present, poor adductors</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

differences between the groups based on socioeconomic status of the parents ($\chi^2 = 7.20; df = 9; p = NS$), access to tertiary-level medical care ($\chi^2 = 11.28; df = 6; p = NS$) or education ($\chi^2 = 8.17; df = 9; p = NS$). Children in the study sample, all of whom had an IQ of 70 or greater, came from higher socioeconomic levels than did the children with an IQ lower than 70 who were omitted from the study ($\chi^2 = 12.13; df = 3; p < .01$). Intelligence quotients below 70 were too few and scattered to allow analysis.

Data Collection

The purpose of the study and the inventory were explained to each parent or guardian whose child met the eligibility criteria. Those willing to participate were instructed first to review the inventory and then to mark with a check all the items their child had achieved. The actual age of a child when he had passed an item was recorded if it had been achieved within the previous four months or if records had been kept in a child's development book. Criteria for all of the items recorded by the parents were reviewed with them and data meeting criteria were transferred to a computer log (CLINFO—Clinical Information System). Parents then were asked to record the exact age at which their child achieved the remaining items over the next 2½ years. Parents were interviewed by phone or in the clinic at three-month intervals, at which time new accomplishments for each child were reviewed, and those items meeting the criteria were transferred to the data pool. The same investigator (JCS) performed all interviews. She was well-acquainted with the majority of children and families in the study and evaluated children when she saw them in the clinic. Participants were dropped from the study if reports disagreed with the results of the clinical evaluation of the child's function.

Parent reports also were evaluated midway through the study. Thirty-three children, a random sample of approximately 20 percent of the study sample, were assessed by professionals not involved with the study but who were working individually with the children. They included community physical therapists and occupational therapists, educators, public health nurses, and psychologists. They were asked to record the child's current function on each inventory item with the response yes, no, or not applicable, using the same criteria used by parents. These reports drew from firsthand observations of the child and from best estimates on items usually confined to the home. The data from parents were then converted to a yes or no format. When the data of the parental and the professional reports were tested for agreement, the concordance was .89. For those items with discordant reports, the parents reported the child was doing the item 66 percent of the time, whereas the professionals reported in the affirmative only 33 percent of the

TABLE 2
Number of Subjects, Sex, IQ, and Ages by Group

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Group 1</th>
<th>Group 2</th>
<th>Group 3</th>
<th>Group 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of subjects</td>
<td>36</td>
<td>33</td>
<td>43</td>
<td>61</td>
</tr>
<tr>
<td>Sex (M/F)</td>
<td>16/20</td>
<td>13/20</td>
<td>24/19</td>
<td>33/28</td>
</tr>
<tr>
<td>IQ*</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>$\bar{X}$ (8)</td>
<td>87.1 (14.3)</td>
<td>94.4 (14.8)</td>
<td>92.1 (10.8)</td>
<td>94.1 (14.3)</td>
</tr>
<tr>
<td>Age (yrs)</td>
<td>7.6 (4.9)</td>
<td>7.3 (5.4)</td>
<td>6.4 (4.9)</td>
<td>6.8 (4.8)</td>
</tr>
<tr>
<td>Age range (yrs)</td>
<td>0.4 - 17</td>
<td>0.3 - 16.7</td>
<td>0.25 - 19.8</td>
<td>0.25 - 17.8</td>
</tr>
</tbody>
</table>

* Includes only children age 3 years and older.
time. Of special note is that there was no difference in concordance between items observed firsthand and those estimated by professionals when compared with parents’ reports. This suggests professionals on the whole had a good understanding of each child’s actual functioning, even when the child was away from a clinic situation.

At the completion of the study, 24 of the 166 items achieved by the children were randomly selected to determine if there were significant differences between items that had been achieved and recorded by parents before the child’s entry into the study (and transferred to the inventory later) and items that had been achieved and recorded during the time of the study. The data from these items were placed in one of two groups, and the mean ages for the groups were calculated. A two-tailed t test showed no differences between them (t = 0.16; df = 46; p = NS). Current and retrospective data were then analyzed together.

Analysis of Data

The percentage of children who passed each item at selected ages was calculated. The age points chosen were at three-month intervals from birth to 4 years and at one-year intervals from 4 to 19 years. The finer analysis was necessary to determine age ranges for the achievement of items that were passed by most children early and within a year’s time. A child was counted at several age points in calculating the percentage of achievement for any given item based on the following criteria: 1) if the child did not pass an item by the end of the study period, he was counted as a no at each age point he crossed during the study period; 2) if the child was able to pass the item at the time of entry into the study but the specific age of passing was unknown, he was counted as a yes at his age at the time of entry into the study but the specific age of passing was unknown, he was counted as a yes at each age point crossed during the study period; and 3) if the specific age at which the child passed the item was known (retrospective or current), he was counted as a yes for that item from the given age to his age at the
end of the study and as a no from birth up to the age
the item was passed. Only in this last case were data
recorded from birth. The age points for 20, 50, and
80 percent achievement for those children for whom
information was known on a given item were selected
to summarize the data. Therefore, the number of
children varied for each of those percentages and for
each of the items.

Criteria chosen to define differences between first
and routine performances reflect the more rapid rate
of development for those items achieved by the
younger age groups. A difference was considered
present 1) if there was a difference greater than 3
months between first and routine performance for
items achieved between the ages of 3 and 24 months,
2) if there was a difference greater than 6 months for
items achieved between the ages of 25 and 72 months,
or 3) if there was a difference greater than 12 months
for items achieved after the age of 72 months.

**RESULTS**

The number of children and the ages at which they
routinely achieved 47 of the 83 items in the eating,
bodily functions, when he asks about his own handi­
items. Three exceptions are in the personal-social
ment for the children who routinely performed the
intermediate steps of sequential tasks. These data
excluding many of the items representing smaller
cover the entire age range of the children in the study
presented in Tables 3 to 6. The items reported here
achievement is applicable: when the child asks about

obvious for items attained after two years of age and
as self-care tasks become more complex. Differences
between groups for the personal-social items are not
so evident because of the variability within the
groups.

Items for which the age ranges of first achievement
and routine performance are different based on cri­
teria described above are indicated in Tables 3 to 6. Children
in Group 1 had significantly more differences
between first and routine performance in all
self-care categories than did any of the other groups
($\chi^2 = 9.32; df = 3; p < .05$). There were no significant
differences among Groups 2, 3, and 4 ($\chi^2 = 1.91; df$
$= 2; p = NS). When categories were compared for
differences between first and routine performance,
there were more differences in the dressing category
($\chi^2 = 35.28; df = 3; p < .001$) than in any other
category ($\chi^2 = 2.56; df = 2; p = NS$). There were only
two items achieved before two years of age for which
there was a difference between first and routine per­
formance (feeds self finger foods, Group 2; takes
socks off, all groups).

**DISCUSSION**

Children with myelodysplasia develop skills in a
sequence comparable to children without handicaps.

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**TABLE 5**

<table>
<thead>
<tr>
<th>Grooming: Numbers of Children and Ages in Months of 20, 50, and 80 Percent Routine Achievement of Selected Items</th>
</tr>
</thead>
<tbody>
<tr>
<td>Item</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>Washes own hands with help getting to sink</td>
</tr>
<tr>
<td>Washes hands without help</td>
</tr>
<tr>
<td>Washes face with help</td>
</tr>
<tr>
<td>Washes face without help</td>
</tr>
<tr>
<td>Assists with brushing teeth</td>
</tr>
<tr>
<td>Brushes teeth without help</td>
</tr>
<tr>
<td>Bathes by self, draws own water</td>
</tr>
<tr>
<td>Dries self after bath, without help</td>
</tr>
</tbody>
</table>

$^a$ Indicates items where criteria were met for a significant difference between the first time the item was accomplished and when it was accomplished routinely.

$^b$ ( ) = n; single. ( ) means each percentage shown had same number of subjects.
<table>
<thead>
<tr>
<th>Item</th>
<th>Group 1</th>
<th>Group 2</th>
<th>Group 3</th>
<th>Group 4</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>20</td>
<td>50</td>
<td>80</td>
<td>20</td>
</tr>
<tr>
<td>Waves “bye-bye”</td>
<td>7</td>
<td>8</td>
<td>11</td>
<td>7</td>
</tr>
<tr>
<td>Is left with babysitter at least once every two months</td>
<td>3</td>
<td>6</td>
<td>24</td>
<td>4</td>
</tr>
<tr>
<td>Explores out of doors</td>
<td>15</td>
<td>24</td>
<td>41</td>
<td>11</td>
</tr>
<tr>
<td>Knows own and others' gender</td>
<td>29</td>
<td>37</td>
<td>39</td>
<td>28</td>
</tr>
<tr>
<td>Helps do small jobs, eg, picks up toys</td>
<td>25</td>
<td>45</td>
<td>51</td>
<td>30</td>
</tr>
<tr>
<td>Participates in simple group games with other children</td>
<td>25</td>
<td>30</td>
<td>46</td>
<td>15</td>
</tr>
<tr>
<td>Knows meaning of and uses &quot;please&quot; and &quot;thank you&quot;</td>
<td>17</td>
<td>22</td>
<td>33</td>
<td>17</td>
</tr>
<tr>
<td>Asks about routine bodily functions</td>
<td>46</td>
<td>53</td>
<td>85</td>
<td>41</td>
</tr>
<tr>
<td>Asks about own handicap</td>
<td>33</td>
<td>48</td>
<td>90</td>
<td>35</td>
</tr>
<tr>
<td>Plays in own neighborhood without being watched</td>
<td>48</td>
<td>78</td>
<td>108</td>
<td>59</td>
</tr>
<tr>
<td>Crosses street without supervision</td>
<td>105</td>
<td>120</td>
<td>120</td>
<td>58</td>
</tr>
<tr>
<td>Goes to friend's house to play</td>
<td>55</td>
<td>69</td>
<td>121</td>
<td>47</td>
</tr>
<tr>
<td>Responsible for routine chores, may be reminded</td>
<td>58</td>
<td>79</td>
<td>72</td>
<td>94</td>
</tr>
<tr>
<td>Paid for working outside home, at least two occasions</td>
<td>92</td>
<td>120</td>
<td>86</td>
<td>186</td>
</tr>
<tr>
<td>Is a member of an organized group</td>
<td>94</td>
<td>132</td>
<td>80</td>
<td>100</td>
</tr>
<tr>
<td>Carries own money and is responsible for making change</td>
<td>110</td>
<td>120</td>
<td>158</td>
<td>67</td>
</tr>
</tbody>
</table>

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* Indicates items where criteria were met for a significant difference between the first time the item was accomplished and when it was accomplished routinely.

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a ( ) = n; single ( ) means each percentage shown had same number of subjects.

b First time only recorded.

c — = the percentage was not reached.

d Indicates items where criteria were met for a significant difference between the first time the item was accomplished and when it was accomplished routinely.
and at a rate dependent in part on the level of their motor paralysis. The wide ranges of achievement within motor-level groups suggest that other factors contribute to delayed development. Children with myelodysplasia have varying degrees of perceptual motor impairment, upper extremity incoordination, scoliosis, kyphosis, and lower extremity contractures. Any one of these factors or the process of their medical management can interfere with the rate of acquiring independence in self-care. The variability of intelligence also may account for the wide range of achievement within groups. We limited our study sample to those with IQs of 70 and above to help minimize this effect. Finer analysis of the variables would require larger numbers. However, many of the children in our study sample with IQs close to 70 were developing at a rate comparable with or even faster than their more intelligent peers.

Sex, social class, race, and environment have been shown to affect achievement in nonhandicapped school-age children but these factors reportedly have little effect before age 1½. Although these variables were not different for the subgroups, they may account for some of the variability within the groups and some of the differences between first and routine performance that were more apparent for the tasks performed at an older age.

Differences in parental expectation and priorities are difficult to test, but these differences, no doubt, account for some of the variability of achievement among similarly paralyzed toddlers and school-age children in this study. For example, parents began to involve their children in self-care of the upper extremities (e.g., washing hands and face with help; assisting with brushing teeth) at approximately the same ages, regardless of the child’s degree of motor paralysis. Yet the ages of the children when parents began teaching lower extremity self-care tasks (e.g., untying shoes, putting on socks with help) were related directly to the degree of their paralysis. Apparently parents have “normal” expectations regarding their child’s capability of performing upper-extremity tasks but overlook the importance of, or postpone teaching methods to deal with, paralyzed lower extremities.

Clearly, this demonstrates a need to provide parents with objective and realistic expectations.

The philosophy of care under which a child receives his medical management also may be expected to affect developmental outcome. Some of the children in the study moved into the clinic area after receiving care elsewhere, others were followed at the clinic from birth. All were recipients of care subject to changing philosophies and improved medical management.

The differences between the ages of first and routine performance may therefore be due to any one or a combination of the factors responsible for within group variability. That these differences were most significant for the most paralyzed group and in the dressing category suggests that time and energy required for routine performance of a task delay the attainment of functional independence. The considerable effort and time required for a paralyzed child to complete a task may preclude his doing it independently at every opportunity if the child must fit into the family routine. It is understandable that rushed parents sometimes quickly complete a task rather than stress independence and also that this assistance may become an expected way of life for the child.

Many of the items showing differences between first and routine performance are those requiring that the parent give the child time and responsibility to perform a task requiring skill or judgment (e.g., using a knife, crossing the street).

The criteria used in this study for routine performance of an item were probably less lenient than criteria used for developmental norms for children without handicaps. In fact, developmental norms for nonhandicapped children do not distinguish between first and routine performance. Yet, routine independent performance of a task contributes to self-reliance.

Some methodological concerns with the design of this study may be raised. Accuracy of parental reporting has been questioned in the past. After reviewing the literature, Hart et al concluded that mothers’ memories become less accurate with time and that they underestimate the age of achieving milestones. However, they found that mothers could provide accurate developmental histories if they were educated about the importance of daily events and recorded them as they occurred. Others have reported mothers’ observations of their children’s daily play and self-care activities to be exceptionally accurate and reliable, and that important milestones were remembered well when parents believed their child was delayed. Our goal was to document daily and routine function and not what the child was capable of doing during a test situation. For this reason, the mothers or the primary care givers were logical record-keepers. The 89 percent concordance between parents and professionals partially validates the accuracy of parent recording.

The number of children in this study is too small to allow further division into more homogeneous subgroups based on intelligence or on detailed variations in motor level of paralysis, associated problems, and demographic factors. Longer periods of data collecting would increase the sample size but might also confound results by introducing such variables as advancing technology and medical management that change over time. A larger sample could be obtained by involving multiple centers and would result in more reliable data, especially for some of the later-achieved, more difficult items.
Pending validation at other regional centers, these guidelines represent the only data describing the rate of achievement of children with myelodysplasia. They provide an objective tool to assess a child's development based on past achievement of similarly disabled children. Once a child's strengths and weaknesses are assessed, parents and therapists can collaborate on possible reasons for a low score, and adaptations in the home environment and therapy program can be initiated. Parents who may be discouraged or reluctant to expect too much too soon of their paralyzed child may be helped to know the earliest possible ages at which important self-care tasks can be attained. It is hoped that further interest in the compilation of data such as these will result in more refined guidelines for children with myelodysplasia.

**CONCLUSION**

The sequence of development of self-care and social tasks by congenitally paralyzed children follows a normal pattern. The rate apparently is dependent not only on the degree of paralysis but also on other physical, social, educational, and medical factors. Complete and systematic documentation of development and associated variables provides the necessary data to study and identify those factors. Then the most effective therapeutic and anticipatory programs can be initiated.

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