The term spina bifida refers to a failure of mesodermal closure around the neural canal. Failure in fusion of neural fold between the twenty-second and the twenty-eighth days of gestation prevents the posterior vertebral arch and the overlying soft tissue from normal development. The spinal cord as well as meningeal structure may herniate out of the bony defect and cause a myelomeningocele, so called spina bifida.

Background. It is controversial to predict ambulation potential using neurological level or specific pattern of muscle strength in patients with spina bifida. We attempted to analyze the relationship between the mobility outcome and its influencing factors utilizing electromyography to study motor unit activity in lower limbs. A subsidiary aim was to evaluate the external anal sphincter with electromyography and to determine the relevant factors of bowel and bladder functions.

Methods. Among 156 patients with spina bifida who had received electromyographic examination and were followed at the out-patient clinic between 1998 and 2000, functional assessment with Pediatric Evaluation of Disability Inventory (PEDI) was applied in 47 randomly selected patients. Other assessments included clinical neurological examination, bowel and bladder function survey, and ambulation evaluation. We correlated the parameters of electromyography with the ambulatory ability and PEDI scores. The innervation of external sphincter was compared between groups with or without neurogenic bowel or bladder dysfunction.

Results. The innervations of hip adductor and quadriceps were found to correlate with walking ability ($p < 0.01$); above muscles as well as anterior tibialis, and gastrocnemius were related to PEDI scores ($p < 0.01$). The neurological level still manifested correlation with walking ability ($p < 0.05$) and PEDI scores ($p < 0.01$). Both denervation potentials and recruitment pattern were important parameters for ambulation and mobility prediction. Significant difference was obtained in denervation potentials of sphincter within bowel groups ($p = 0.036$) and bladder groups ($p = 0.016$).

Conclusions. Both traditional neurological level and specific muscle innervation exert crucial influence on walking and mobility functions. Electromyographic assessment demonstrates its contribution in prediction of functional outcome in spina bifida.

[Chin Med J (Taipei) 2001; 64: 509-515]

Received: March 13, 2001. Accepted: June 22, 2001.
Correspondence to: Rai-Chi Chan, MD, Department of Physical Medicine and Rehabilitation, Taipei Veterans General Hospital, 201, Sec 2, Shih-Pai Road, Taipei 112, Taiwan. Fax: +886-2-2875-7359; E-mail: rcchan@vghtpe.gov.tw
bifida aperta. Spina bifida occulta decribes a pos terior bony deect with out men in geal or neur al el ements in volve ment. Lipomyelomeningocele is lipomas derived from epider mal and mesodermal tis sue within the neur al ca nal. They grow through the ver te bral deect and at tach to an el on gated, teth er ed spi nal cord. The pres sure or ad he sion ef ect in the ca nal of ten re sults in meningocele for ma tion and ab nor mal neur al development.

The clin i cal im por tance of the spina bifida is not only the ab nor mal ity of the spine, but the in volve ment of the cen tral ner vous sys tem, musculoskeletal com pli ca tions, neurogenic bowel and blad der, and psy-cho social is sues. The level of neu ro log i cal le sion and the pat tern of lower-extremities innerva tion are the most de ter mi nant fac tors in flu encing whether chil dren with spi nal bifida at tain func tional ab il ity. How ever, pre vi ous au thors have also found the im por tance of am bu la tion of quad riceps, hip ab ductors, and knee flex ors. A sys tem atic study at tempt ed to clar ify am bu la tion po ten tial by study ing se rial man ual mus cle test ing in iliopsoas, quad riceps and tibialis an te rior mus cles in stead of study ing neu ro log i cal ex am i na tion. We re viewed the med i cal charts of 156 pa tients who had re ceived EMG stud ies and were fol low ed at the spina bifida clinic of Tai pei Vet erans Gen eral Hos pital be tween 1998 and 2000. All chil dren un der went sur gical treat ment of spi nal cord mal for ma tion early after birth and were in stable clinical condi tion. Electromyography (EMG) was per formed in each pa tient at a mean age of 1 year via a Nicolet Vi king II or Key point re cording ap par a tus with a stan dard con cen tric needle Electrode. Motor unit ac tiv ities in hip adductor, quad riceps, an ter ior tibialis, gas trocneu mius, ex ter nal anal sphinctor were re cording at rest as well as during vol i tional con tra ction. The re cord ing para me ters in cluded the denerva tion po ten tials (pos i tive sharp waves, fi bril la tions), re cruit ment pat tern and the neu ro log i cal le vel. The denervation po ten tials were graded as 1-5 ac cord ing to their num ber and ease of ob serv ation based upon the num ber of sites ex am ined (Table 1).

The po ten tial pat tern was graded as 1-6 (Table 1). Ac cording to neu ro log i cal ex am i na tion, we cate gORIZED the clin i cal neu ro log i cal le vel as tho racic, high-lumbar (L1, 2), mid-lum bar (L3), lumbos ac ral (L4, 5-S1), and lower-sacral level (S2-4). The EMG lo cal iza tion of neu ro log i cal le vel had the same clas si fi ca tion. The EMG lo cal iza tion of neu ro log i cal le vel had the same clas si fi ca tion.

**Table 1. Grading of denervation potentials and recruitment patterns**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Denervation potentials</th>
<th>Recruitment patterns</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>No Fib/PSW potentials</td>
<td>RR ≤ 5</td>
</tr>
<tr>
<td>2</td>
<td>Persistent/unsustained single trains in at least two muscle regions</td>
<td>5 &lt; RR ≤ 10</td>
</tr>
<tr>
<td>3</td>
<td>Moderate numbers in three or more muscle areas</td>
<td>10 &lt; RR ≤ 20</td>
</tr>
<tr>
<td>4</td>
<td>Many in all muscle regions</td>
<td>20 &lt; RR ≤ 30</td>
</tr>
<tr>
<td>5</td>
<td>Baseline obliterated with Fib/PSW in all areas of muscles examined</td>
<td>RR &gt; 30</td>
</tr>
<tr>
<td>6</td>
<td>No motor unit potential</td>
<td></td>
</tr>
</tbody>
</table>

Fib = fibrillation potentials; PSW = positive sharp waves; RR = recruitment ratio = Firing rate (Fastest)/Number of motor unit potentials. ° Modified from Dumitru D, Electrodiagnostic Medicine. 22
tion. Raw scores for functional skills and caregiver assistance are converted to normative standard and scaled scores. We utilized the mobility scaled scores in functional skill and caregiver assistance dimensions for correlation study. Forty-seven patients were randomly selected from the 156 patients for administration of PEDI, including 15 myelomeningocele and 32 lipomyelomeningocele with out the involvement of upper motor neuron. Twenty-seven boys and twenty girls were enrolled in the cross sectional study, with mean age of 4.4 years (ranged from 0.5 to 12 years). The interview with parents or guardians of each child was scheduled for 40-50 minutes and conducted by the author. Walking ability was also evaluated in children older than 2 years and graded as independent ambulator, partially ambulatory, and non-ambulatory. Independent ambulation was defined as walking with or without ankle-foot orthoses. Partial ambulation referred to walking with crutches or walkers. Children who scooted or crawled, or used a stroller or wheelchair for locomotion were defined as being non-ambulatory.

Patients who had fecal incontinence, constipation or anal mal tone were considered to have neurogenic bowel. Voiding disorders combined with the anal mal finding in sonography or urodynamic study was taken into account for neurogenic bladder. We divided all subjects into normal bowel group vs. neurogenic bowel group for the study of bowel function, and also divided them into normal bladder group vs. neurogenic bladder group for study of bladder function.

To determine the most important lower extremity muscles being involved in the mobility of PEDI in each muscle with PEDI mobility scaled scores and walking ability using Spearman rank correlation analysis. Parameters of EMG in external sphincters were compared between bowel and bladder groups using Chi-square test.

### Results

The clinical characteristics of our subjects are listed in Table 2. The neurological level determined by electromyography (ENL) showed high correlation with clinical neurological level (CNL). Regarding walking ability, 77% of patients walked independently, 4.2% were partially ambulatory, and 19% were non-ambulatory. There were 55% and 68% of patients found to be with neurogenic bowel or bladder, respectively.

The correlations of EMG parameters with walking ability and PEDI mobility scaled scores are presented in Table 3. The key muscles important to ambulation ability were hip adductor and quadriceps. The grading of denervation potentials in quadriceps as well as recruitment pattern in adductor and quadriceps showed high correlation with walking ability. Recruitment in adductor and quadriceps showed significant difference (p = 0.036) was obtained only in denervation of external sphincter be tween bowel groups. Also, only the denervation of external sphincter was significantly different (p = 0.016) between bladder groups (Table 4).

### Table 2. Summary of clinical characteristics

<table>
<thead>
<tr>
<th>Neurological level</th>
<th>Case numbers (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>No deficit</td>
<td>CNL 13 (27.7%)</td>
</tr>
<tr>
<td>Lower-sacral (S2-4)</td>
<td>12 (25.5%)</td>
</tr>
<tr>
<td>Lumbosacral (L4-S1)</td>
<td>19 (40.4%)</td>
</tr>
<tr>
<td>Mid-lumbar (L3)</td>
<td>1 (2.1%)</td>
</tr>
<tr>
<td>High-lumbar (L1-2)</td>
<td>1 (2.1%)</td>
</tr>
<tr>
<td>Thoracic level</td>
<td>1 (2.1%)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Walking ability</th>
<th>Case numbers (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Independent</td>
<td>36 (76.6%)</td>
</tr>
<tr>
<td>With assistive device</td>
<td>2 (4.2%)</td>
</tr>
<tr>
<td>Non-ambulatory</td>
<td>9 (19.1%)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Bowel groups</th>
<th>Case numbers (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal bowel function</td>
<td>21 (44.7%)</td>
</tr>
<tr>
<td>Neurogenic bowel</td>
<td>26 (55.3%)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Bladder groups</th>
<th>Case numbers (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal bladder function</td>
<td>15 (31.9%)</td>
</tr>
<tr>
<td>Neurogenic bladder</td>
<td>32 (68.1%)</td>
</tr>
</tbody>
</table>

CNL = clinical neurological level; ENL = electromyography determined neurological level.

* Spearman rank correlation between ENL and CNL is significant at the 0.01 level.
Discussion

The actual pattern of voluntary motor innervation, that is, the clinical neurological level, is best determined by careful inspection of muscle bulk, stimulation with posture challenges and gentle opposition of major muscle groups to elicit voluntary activity. Clinically defined pattern of muscle strength, according to neurosegmental motor level, was firstly proposed by Sharrard in 1964.\textsuperscript{16} Traditionally, the motor level is considered to be the most determinative factor in predicting ambulation potential.\textsuperscript{3,4} The higher the motor level, the more problems such as joint contracture, spinal and hip deformity that should be prevented if possible.\textsuperscript{17,18}

However, McDonald et al. found that the patterns of muscle strength of ten of the classic neurosegmental pattern in myelomeningocele.\textsuperscript{19} Samuelsson and Skoog announced that the large disparity in ambulatory status among patients with similar patterns of neurosegmental innervation may be due to the use of broad categories proposed by Sharrard.\textsuperscript{7} Previous studies had stressed the importance of iliopsoas, quadriceps, hip abductors, and knee flexors strength in prediction of ambulation potential.\textsuperscript{8-11} The purpose of our study is to identify the most influencing factors in mobility development of spina bifida.

With regard to the investigative method, we used EMG instead of manual muscle testing because of several advantages. First, unlike the application of EMG without limitation of patient’s age, the manual muscle testing is better administered in children older than 5 years. Previous authors pointed out that the perceived accuracy of manual muscle testing peaks at the age of five and the muscle strength will remain stable after that age.\textsuperscript{20} The estimation of mobility potential can be attained in early period after childbirth by EMG study. When we approach an infant, muscle relaxation to examine spineous or abnormally inser tional ac tivity is obtained by placing the muscle at its shortest end from its own sition. Re cruitment of motor units in an infant is often difficult to interpret but can be facilitated by primitive reflex or withdrawal response. Besides, the muscle present of re cruit-

\begin{table}
\centering
\caption{The correlative coefficient ($r$) between EMG parameters and mobility scores}
\begin{tabular}{lll}
\hline
 & Walking ability & Functional skill score & Caregiver assistance score \\
\hline
Hip adductor & 0.311 & 0.268 & 0.283 \\
Denervation & 0.481\textsuperscript{a} & 0.342\textsuperscript{b} & 0.338\textsuperscript{b} \\
Recruitment & & & \\
Quadriceps & 0.446\textsuperscript{a} & 0.330 & 0.359\textsuperscript{b} \\
Denervation & 0.489\textsuperscript{a} & 0.397\textsuperscript{a} & 0.421\textsuperscript{a} \\
Recruitment & & & \\
Anterior tibialis & 0.322 & 0.394\textsuperscript{a} & 0.412\textsuperscript{a} \\
Denervation & 0.285 & 0.447\textsuperscript{a} & 0.415\textsuperscript{a} \\
Recruitment & & & \\
Gastrocnemius & 0.301 & 0.420\textsuperscript{a} & 0.434\textsuperscript{a} \\
Denervation & 0.277 & 0.374\textsuperscript{b} & 0.410\textsuperscript{a} \\
Recruitment & 0.349\textsuperscript{b} & 0.500\textsuperscript{a} & 0.518\textsuperscript{a} \\
ENL & & & \\
\hline
\end{tabular}
\footnotesize{ENL = electromyography determined neurological level.}
\footnotesize{\textsuperscript{a}Spearman rank correlation is significant at the 0.01 level.}
\footnotesize{\textsuperscript{b}Spearman rank correlation is significant at the 0.05 level.}
\end{table}

\begin{table}
\centering
\caption{The $p$-value in comparing the EMG parameters of external sphincter in the bowel and bladder groups}
\begin{tabular}{lll}
\hline
 & Denervative potentials & Recruitment patterns \\
\hline
Bowel groups & 0.036\textsuperscript{a} & 0.639 \\
Bladder groups & 0.016\textsuperscript{a} & 0.071 \\
\hline
\end{tabular}
\footnotesize{\textsuperscript{a}Chi-square test revealed significant difference at $p < 0.05$.}
\end{table}
ment ratio does not require maximal contraction; conversely, it needs minimal to moderate muscle contraction. Secondly, isolated muscle was approached in the EMG study, unlike the manual muscle testing in which the recording of summative strength in a functional muscle group is usually in evitable, especially when the subject is a child. Thirdly, the inter pretation of the bioelectric activity is quantified on the EMG recorder oscilloscope, but the judgment of perceived muscle power is usually subjective. Finally, the neurologic level determined by EMG study also showed high correlation with clinical neurologic level. This finding confirms the reliability of the EMG determined neurologic level.

We recommend the use of both neurologic and specific patterns of lower extremity strength for predicting mobility and walking ability. EMG neurologic level is an important factor influencing functional outcome, in spite of the variable correlation between the classic neurosegmental levels and the motor pattern in McDonald's observation. The level of neurologic impairment determined by EMG study also showed high correlation with clinical neurologic level. This finding confirms the reliability of the EMG determined neurologic level.

In summary, the innervation of hip flexor and quadriceps is crucial for walking ability; that of hip flexor, quadriceps, and tibialis anterior is essential for quality of mobility. Neurologic level still has a significant influence on walking ability and mobility. Both denervation potential and recruitment patterns should be taken into account in assessing potential outcome in children with spine bifida. In conclusion, EMG studies in lower limbs and sphincter muscles can contribute to the prediction of functional outcome in patients with spina bifida.

References

肌電圖檢查應用於脊柱裂患者預後的評估

蔡泊意1 詹瑞棋1,3 楊翠芬1,3 黃棣棟2,3 黃苾忻1 潘柏榮1

台北榮民總醫院 復健醫學部1 神經醫學中心小兒神經外科2
國立陽明大學 醫學院3

背景 對於脊柱裂患者之行走能力，使用神經功能位置的高低或下肢肌肉強度作為預後之預測仍有爭論。本研究利用肌電圖檢查評估行動能力與其影響因子之間的關係，並探討肛門外擴約肌之肌電圖參數與排便及膀胱功能的關係。

方法 研究對象包括自民國八十七年至民國八十九年於本院定期追蹤及隨機取得之四十七位脊柱裂患者，每位病患均接受神經學檢查、膀胱功能檢查、肌電圖檢查及兒童功能障礙之評估（PEDI）。將肌電圖檢查之結果與行走能力及 PEDI 之分數做相關測試，並比較外擴約肌之肌電圖檢查結果在受損與未受損排便及膀胱功能之病患的不同。

結果 髖內收肌與股四頭肌和行走能力有顯著相關（p < 0.01）；髖內收肌、股四頭肌、
脛前肌、腓腸肌與活動能力有明顯相關（p < 0.01）。而神經受損位置的高低與行走（p < 0.05）及活動能力（p < 0.01）均有顯著相關。在肌電圖參數方面，去神經現象的多寡及神經元徵召的程度均對行走及活動能力有顯著影響。在排便及膀胱功能受損的病患只有外擴約肌的去神經現象和正常的病患有不同。

結論 傳統的神經功能位置高低與下肢肌肉強度對於行走及活動能力評估均有預測之價值。外擴約肌去神經現象的多寡可做為排便及膀胱功能的預測。

關鍵詞 膀胱功能、排便功能、肌電圖檢查、活動能力、脊柱裂。