



Neurological abnormalities, major orthopaedic deformities and ambulation analysis in a myelomeningocele population in Catalonia (Spain)

J Iborra*¹, E Pagès¹ and A Cuxart¹

¹*Spina Bifida Unit, Hospital de Traumatología y Rehabilitación, Vall d'Hebron Hospitals, Barcelona, Spain*

The aim of the study was to analyze the present status of neurologic abnormalities, major orthopaedic deformities and ambulatory status in a large myelomeningocele population.

Patients and methods: Cross-sectional study based on the clinical and radiographic records of 322 patients treated and followed-up from 1967–1995. The setting was a multidisciplinary spina bifida unit within a third-level university hospital, which serves as the referral centre for these patients in Catalonia (Spain). We collected information on diagnosis, central nervous system, musculoskeletal system (spinal and hip deformities) and functional level in each patient. To study relationships among the variables, the Mann-Whitney U and the Chi-squared tests were applied. Results were considered to be statistically significant at *P* levels of ≤ 0.05 .

Results: Mean age was 15.9 years. 78.1% of patients had mid-lumbar, low-lumbar or sacral neurological levels; 97.5% had hydrocephalus and 68.8% were shunted. Prevalence of spine deformities was 45.3%; 38.8% had dislocation of one or both hips. Median age of walking onset was 37.1 months and 74.8% of patients were ambulatory. Median age at which ambulation ceased was 128 months (10 years and 8 months). The bivariate analysis showed statistically significant relationships between neurological level and all the variables studied ($P < 0.001$, $P < 0.02$) except body mass indexes and intelligence quotient.

Conclusions: Neurological level was the main factor that determined neurological abnormalities, major orthopaedic deformities and ambulatory status.

Keywords: myelomeningocele; neurological abnormalities; orthopaedic deformities; ambulatory status

Introduction

Myelomeningocele (MMC) is a complex syndrome that affects mainly the nervous, musculoskeletal and genitourinary systems. It is the second most frequent cause of infantile disability after cerebral palsy.^{1,2} Over the last 30 years, a great deal of effort has been directed toward the treatment of this disorder.

The currently used methods for treating MMC patients were first introduced in the 1960's. They consist of closure and cutaneous coverage of the myelomeningocele defect by operating in the first days of life, and later, control of hydrocephalus with ventriculoperitoneal cerebrospinal fluid shunting. This therapy, together with improvements in treating the neurogenic bladder, has made it possible for these patients to reach adulthood. Now that long-term survival has been reasonably controlled, attention has been focused on other problems, such as definitive control of the hydrocephalus, musculoskele-

tal problems, urinary and faecal continence, and the education, social integration and economic independence of these patients.^{1–5}

Our aim with this epidemiologic study was to analyze the present situation regarding neurologic abnormalities, major orthopaedic deformities (particularly scoliosis and hip dislocation) and ambulatory status in MMC patients who have been attended at our hospital over the last 30 years.

Methods

We performed a cross-sectional study based on the clinical and radiologic records of patients who were treated and followed-up in a multidisciplinary unit from 1967–1995. Our Spina Bifida Unit is the referral centre for the Catalanian Autonomous Community in Spain. Over the study period, 487 patients with congenital malformation of the neural tube were attended and 394 corresponded to patients with MMC. From this group, 17 who had died over the years and 55 in whom periodic control had been interrupted during the previous 3 years were excluded

* Correspondence: J Iborra, Spina Bifida Unit, Hospital Universitario de Traumatología y Rehabilitación, Vall d'Hebron Hospitals, P Vall d'Hebron 119-139, 08035 Barcelona, Spain

from the study. Thus, 322 MMC patients comprised the final series.

The study protocol was designed specifically for deformities of the spine⁶ and included personal data, general information, diagnosis, data related to nervous system pathology, description of spine deformities and their treatment, data related to hips and treatment, and evaluation of ambulatory status.

Patients were weighed (kg) sitting on a platform scale. Height (cm) was measured in the decubitus position on a rigid surface with an incorporated metric measure. Arm span determination was carried out in a sitting position. To establish the ideal weight-to-height relation, we used the body mass indexes (BMI) described by Quetelet⁷ ($\text{weight}/\text{height}^2 \times 100$) and Roche⁸ ($\text{weight}/\text{corrected arm span}^2 \times 100$). Arm span was corrected according to neurologic level: at the thoracic and high lumbar levels (T10–L2) arm span was multiplied by 0.9, at the middle and low lumbar levels (L3–L5) by 0.95 and at the sacral levels by 1.^{2,8}

Motor examination was carried out separately for the right and left sides. In this work, we always refer to the functional neurologic level (and not the lesion), as defined by the lowest level muscle capable of an antigravitatory force of grade 3 or more on the MRC scale.⁹ Patients were grouped according to functional levels: thoracic (T6–T12), high lumbar (L1–L2), mid-lumbar (L3), low lumbar (L4–L5) and sacral (S1–S5). When relating the different variables to the neurologic level, the most highly affected of the two sides was chosen.

Psychometric study was carried out with the revised Weschler intelligence scale for adults (WISA-R) in patients over 16 years old, and with the scale for children (WISC-R) in patients under 16.¹⁰ Radiological study of the spine included anteroposterior and lateral X-rays with the patient seated. Spine deformities were recorded according to the criteria of the Scoliosis Research Society.¹¹

Passive joint range of motion (flexion and extension) was measured in both hips with the patient on a hard surface in a supine or prone decubitus position. To relate this variable with the others, patients were grouped according to their hip flexion contracture: (1) no flexion contracture in either hip; (2) 25° or more flexion contracture in at least one hip and (3) 25° or more flexion contracture in both hips. Anteroposterior X-rays of the hips in the supine decubitus position were performed in all patients. Hip stability was evaluated using Reimer's migration index.^{12,13} Functional ambulation was defined according to Hoffer's classification.¹⁴ Independent mobility was defined as the capacity for, and effectiveness of, free movement, whether walking or with use of a wheelchair.²

Statistical analysis was performed using the BMDP software package (University of California Press, Berkeley, CA, 1985). An initial descriptive analysis of the data was carried out using the mean, standard deviation and range for quantitative data, and the absolute and relative frequencies corresponding to

each category for the qualitative data. To study the relationships among the different variables, the non-parametric Mann-Whitney U test was applied for quantitative data and the Chi-squared test for qualitative data. Results were considered to be statistically significant at *P* levels of ≤ 0.05 .

Results

The descriptive data of the patients are shown in Tables 1–3. Table 4 shows relationships between body mass index and age. Tables 5–7 show relationships between neurologic level and shunted hydrocephalus, brain lesion, spine deformity, hip flexion-contracture, hip status on radiology, age at the beginning and cessation of ambulation, beginning of independent mobility and ambulatory status. In the bivariate analysis, a statistically significant relationship was found between neurologic level and all the variables

Table 1 Variables related to general information

| | No patients (%) |
|-----------------------------|---|
| <i>Age</i> | |
| 0–5 years | 44 (13.7%) |
| 6–10 years | 50 (15.5%) |
| 11–15 years | 47 (14.6%) |
| 16–20 years | 77 (23.9%) |
| 21–25 years | 75 (23.3%) |
| <25 years | 29 (9.0%) |
| <i>Mean age</i> | 15.9 years (±8; limits 1–48 years) |
| <i>Sex</i> | |
| Males | 177 (55.0%) |
| Females | 145 (45.0%) |
| <i>Menarch (n = 145)</i> | |
| Yes | 101 (70.0%) |
| No | 44 (30.0%) |
| <i>Mean age menarche</i> | 11.2 years (±1.4; limits 9–16 years) |
| <i>Body mass indices</i> | |
| <i>Quetelet index</i> | |
| <22 (normal) | 133 (41.3%) |
| 22–25 (over-weight) | 51 (15.8%) |
| 26–30 (obesity) | 75 (23.3%) |
| >30 (morbidly obesity) | 63 (19.6%) |
| <i>Roche index</i> | |
| <0.22 (normal) | 106 (32.9%) |
| 0.22–0.25 (over-weight) | 91 (28.3%) |
| 0.26–0.30 (obesity) | 87 (27.0%) |
| >0.30 (morbidly obesity) | 38 (11.8%) |
| <i>*Mean Quetelet index</i> | |
| Males | 23.9 (±6.3; limits 14.1–44.4) |
| Females | 23.7 (±5.9; limits 13.7–38.9) |
| <i>*Mean Roche index</i> | |
| Males | 0.24 (±0.06; limits 0.15–0.42) |
| Females | 0.25 (±0.06; limits 0.14–0.42) |

studied, except body mass indexes and Intelligence Quotient (IQ).

When the brain lesion and IQ variables were evaluated together, there was a statistically significant relationship between presence of a brain lesion and IQ: 70% of patients with brain lesions had low IQs ($P < 0.0001$).

Discussion

The largest number of patients in our series was born in the decade of 1969–1978 (45.3%), a time of demographic increase in Catalonia.¹⁵ They were the first generation of MMC patients in our setting to

Table 2 Variables related to neurological level

| | Right side | Left side | No of patients |
|--|------------|-----------|----------------|
| <i>Neurological level</i> | | | |
| Thoracic (T10–T12) | 31 | 35 | |
| High lumbar (L1–L2) | 34 | 33 | |
| Mid lumbar (L3) | 79 | 74 | |
| Low lumbar (L4–L5) | 120 | 118 | |
| Sacral (S1–S5) | 58 | 62 | |
| <i>Functional neurological level</i> | | | |
| Thoracic (T10–T12) | | | 36 (11.2%) |
| High lumbar (L1–L2) | | | 34 (10.5%) |
| Mid lumbar (L3) | | | 77 (23.9%) |
| Lumbar (L4–L5) | | | 119 (36.9%) |
| Sacral (S1–S5) | | | 56 (17.3%) |
| <i>Neurological level</i> | | | |
| Symmetrical | | | 294 (91.3%) |
| Asymmetrical (1) | | | 20 (6.2%) |
| Asymmetrical (2) | | | 8 (2.5%) |
| <i>Hydrocephalus</i> | | | |
| Yes | | | 314 (97.5%) |
| No | | | 8 (2.5%) |
| <i>Shunted hydrocephalus (n = 314)</i> | | | |
| Yes | | | 216 (68.8%) |
| No | | | 98 (31.2%) |
| <i>Brain lesion</i> | | | |
| Yes | | | 57 (17.7%) |
| No | | | 265 (82.3%) |
| <i>Intelligence quotient</i> | | | |
| Normal (80–130) | | | 141 (43.8%) |
| Borderline (70–79) | | | 48 (14.9%) |
| Low (≥ 69) | | | 133 (41.3%) |
| <i>Normal shunt function (n = 216)</i> | | | |
| Yes | | | 206 (95.4%) |
| No | | | 16 (4.6%) |
| <i>Other neurological alterations</i> | | | |
| Yes, trunk | | | 137 (42.5%) |
| Yes, some of several | | | 30 (9.0%) |
| No | | | 155 (48.1%) |

Table 3 Variables related to hips and spine

| | Right side | Left side | No of patients |
|---|------------|-----------|----------------|
| <i>Spine deformity (n = 322)</i> | | | |
| Yes | | | 146 (45.3%) |
| No | | | 176 (54.7%) |
| <i>Type of spine deformity (n = 146)</i> | | | |
| Scoliosis | | | 131 (89.7%) |
| Kyphosis | | | 15 (10.3%) |
| <i>Etiology of spine deformity (n = 146)</i> | | | |
| Congenital | | | 10 (6.9%) |
| Paralytic | | | 96 (65.7%) |
| Idiopathic | | | 40 (27.4%) |
| <i>Osseous rachischisis (n = 322)</i> | | | |
| T12 | | | 26 (8.1%) |
| L1 | | | 26 (8.1%) |
| L2 | | | 30 (9.3%) |
| L3 | | | 77 (23.9%) |
| L4 | | | 96 (39.8%) |
| L5 | | | 50 (15.5%) |
| Sacral (S1–S5) | | | 17 (5.2%) |
| <i>Pelvic obliquity (n = 322)</i> | | | |
| Yes, R > L | | | 74 (23%) |
| Yes, L > R | | | 57 (17.7%) |
| No | | | 191 (59.3%) |
| <i>Length leg discrepancy (n = 322)</i> | | | |
| Yes, R > L | | | 75 (23.3%) |
| Yes, L > R | | | 85 (26.4%) |
| No | | | 162 (50.3%) |
| <i>Stable sitting function (n = 322)</i> | | | |
| Yes | | | 309 (96%) |
| No | | | 13 (4%) |
| <i>Treatment for spine deformity (n = 146)</i> | | | |
| Surgery | | | 26 (17.8%) |
| Conservative with body brace | | | 20 (13.7%) |
| Observation | | | 100 (68.5%) |
| <i>Hip flexion contracture $\geq 25^\circ$</i> | | | |
| Yes, one hip | | | 32 (9.9%) |
| Yes, both hips | | | 56 (17.4%) |
| No | | | 234 (72.7%) |
| <i>Radiological status of hips</i> | | | |
| Centred | 233 | 214 | |
| 1 Subluxated or dislocated | 25 | 40 | |
| 2 Subluxated or dislocated | 64 | 68 | |
| <i>Radiological status of hips</i> | | | |
| Centred | | | 197 (61.2%) |
| 1 Subluxated or dislocated | | | 53 (16.5%) |
| 2 Subluxated or dislocated | | | 72 (22.4%) |
| <i>Surgery</i> | | | |
| Yes | | | 70 (21.7%) |
| No | | | 252 (78.3%) |

benefit from early closure of the myelomeningocele defect, shunting, and prevention of urological complications. Patients born during the decade of 1979–1988, when demographic pressure had decreased, were the second largest group (30.1%). By that time routine prenatal diagnosis and systematic application of selection criteria had been established. The third group consisted of those born from 1988–1995 (13.7%). The relatively small number of cases under 6 years old is due to the overall decrease in birth rate in our country,¹⁵ the more generalised use of folic acid prophylaxis in fertile women,¹⁶ and the documented tendency toward a generalised decrease in patients with this neural tube defect.^{17–19} The small number of cases born before 1967 corresponds to patients with low neurologic levels who survived spontaneously.

Table 4 Bivariate analysis age–body mass indexes

| Age (years) | Mean Quetelet index | Mean Roche index |
|-------------|---------------------|------------------|
| 0–5 | 18 | 0.2 |
| 6–10 | 19.6 | 0.2 |
| 11–15 | 23.5 | 0.24 |
| 16–20 | 26.9 | 0.26 |
| >20 | 26.1 | 0.27 |
| <i>P</i> | <0.001 | <0.001 |

Distribution by gender showed a male-to-female ratio of 1.2:1, a result that is similar to the 1.1:1 reported by Samuelsson and Eklöf,²⁰ and in contrast to other series that show an inversion of this ratio.^{5,21–24} Mean weight was within the 10th–50th percentiles, and mean height and mean corrected arm span were below the 3rd percentile.²⁵ As expected, males were taller and weighed more than females. According to normal percentile tables, patients were not overweight in absolute terms. However, when the ideal relationship between weight and height or corrected arm span (expressed by the Quetelet and Roche indexes) was analyzed, we observed that the mean for the two indexes was within the margin of overweight. Fifty-nine per cent and 67% of the patients presented Quetelet and Roche indexes, respectively, over the normal values, a finding that has been reported in other studies in which the percentages of patients with above-normal BMIs ranged from 27% to 90%.^{26–28}

Thus, we found that patients with MMC were shorter and weighed more than normal for their ages. Analysis of the BMI according to age group showed normal values up to 10 years of age. At 10 years and over these values increased, and at 15 years and over they reached obesity. Causes of obesity in these patients are multiple. They include lower basal energy requirements and lower energy consumption during daily activity, which is exacerbated by the tendency toward immobility and a sedentary life style after 10 years of age.^{6,26–28}

Table 5 Bivariate analysis of neurological level

| Neurological level | % Patients shunted (n=314) | % Patients with brain lesions (n=322) | % Patients with spine deformity (n=322) | % Patients with 1 or 2 ≥25° hip flexion contracture (n=322) | % Patients with 1 or 2 subluxated or dislocated hips (n=322) |
|--------------------|----------------------------|---------------------------------------|---|---|--|
| ≥T12 | 80 | 30.5 | 85.7 | 61.1 | 69.4 |
| L1–L2 | 90.6 | 38.2 | 61.8 | 55.9 | 76.5 |
| L3 | 74.7 | 15.6 | 55.3 | 41.6 | 84.4 |
| L4–L5 | 66.7 | 8.3 | 36.7 | 10.9 | 22.7 |
| Sacral | 45.5 | 19.6 | 16.1 | 3.6 | 3.6 |
| Total | 69 | 17.7 | 45.3 | 27.3 | 38.8 |
| <i>P</i> | <0.001 | <0.001 | <0.001 | <0.001 | <0.001 |

Table 6 Bivariate analysis of neurological level

| Neurological level | Age (months) | | |
|--------------------|-----------------------------|------------------------------|-----------------------------|
| | Initiation ambulation | Cessation ambulation | Independent mobility |
| ≥T12 | 73 (±19.1; limits 48–108) | 115.3 (±44.4; limits 56–180) | 67.6 (±27.7; limits 16–120) |
| L1–L2 | 47.7 (±16.2; limits 18–72) | 104.3 (±23.4; limits 78–144) | 51.3 (±18.9; limits 8–90) |
| L3 | 46.4 (±35.9; limits 17–216) | 167.1 (±44; limits 72–216) | 54.4 (±35.5; limits 17–216) |
| L4–L5 | 32 (±20.2; limits 10–144) | – | 37.6 (±24.6; limits 13–192) |
| Sacral | 26.4 (±13.7; limits 11–72) | – | 29 (±17.7; limits 11–108) |
| <i>P</i> | <0.02 | <0.02 | <0.02 |

Table 7 Bivariate analysis of neurological level

| Neurologic level | Community (610m) | Household (30m) | Ambulatory status ($P < 0.001$) | | Total |
|------------------|------------------|-----------------|-----------------------------------|----------------|-------|
| | | | Non-functional (< 3 m) | Non-ambulators | |
| ≥ T12 | 1 | 1 | 0 | 34 | 36 |
| L1–L2 | 7 | 4 | 1 | 22 | 34 |
| L3 | 42 | 16 | 4 | 12 | 77 |
| L4–L5 | 103 | 9 | 4 | 3 | 119 |
| Sacral | 54 | 1 | 1 | 0 | 56 |
| Total | 210 | 31 | 10 | 71 | 322 |

The mean age of menarche in our women patients (11.2 years) was lower than in the normal Catalonian population (12.3 years) according to data published by Puente *et al*²⁵ and higher than patients in the series reported by Menelaus⁵ and Shurtleff² (9 years).

In the great majority of cases (91%) neurologic symmetry was observed, a relatively original finding in the context of the literature.⁶ The level of the neurosegmental lesion was predominantly mid-lumbar, low lumbar and sacral, and was similar to distributions reported in other series,^{2,4–6} although there were comparatively fewer patients with thoracic and high lumbar levels. The small differences in the literature are due to the fact that criteria for classifying neurological levels were not uniform.

Hydrocephalus was present in 314 of the 322 patients and 31% had not been shunted. At present, we are only sure that 20 patients of the total with hydrocephalus have evidence of intracranial hypertension, as confirmed by clinical signs and/or continuous intracranial pressure (ICP) monitoring. This figure may increase after the work by Sahuquillo *et al*²⁹ which conclusively demonstrates that there are patients with no clinical signs of intracranial hypertension who have active hydrocephalus and are in need of shunting. Hydrocephalus, is therefore, a problem that cannot be considered definitively resolved. As would be expected, patients in our series with higher neurological levels were the ones most often shunted.

One noteworthy finding was the large percentage of cases with central neurological involvement or brain lesions (18%), as compared to the 7.6% found in the Göteborg series.⁴ These lesions may be sequelae of the hydrocephalus and its treatment, or may be considered a part of the MMC syndrome. When brain lesions were related with neurological level, we observed the same distribution that appeared in the relationship between shunted hydrocephalus and neurological level, except for the large number of patients with sacral level involvement and brain lesions (19.6%). Individual analysis showed a clear relationship between brain lesions and complications caused by hydrocephalus and its treatment.

The IQ was found to be low in the majority, with 56% of patients situated at borderline level or below.

The mean IQ in our study was similar to results reported by Tew and Laurence³⁰ and Anderson and Spain,³¹ and lower than results from Soare and Raimondi³² and McLone *et al*.³³ Once again, these discrepancies can be attributed to differences in criteria (in this case definition of IQ) and to the heterogeneity of the samples. Overall distribution of IQ in our series was not gaussian or normal: the distribution curve deviated toward the left. There was no relationship between neurological level and IQ; however, there was a clear relationship between brain lesion and IQ. Brain lesions, mainly due to problems in diagnosis and treatment of hydrocephalus, were determinant for intellectual capacity in our patients.

Regarding the variable, 'other neurologic abnormalities', we wish to clarify that not all the patients were exhaustively studied in this respect and that the frequency of these anomalies will undoubtedly be found to increase as systematic study with magnetic resonance (MR) techniques is established.³⁴ Systematic MR testing implies considerable expense, and the studies published to date using MR in MMC have been retrospective, not prospective, or motivated by the neurologic deterioration of the patient.

Spine deformities in MMC are usually paralytic and progressive and may cause a worsening of the established disability in these children. This fact can interfere with rehabilitation and can frustrate previous treatments to maintain ambulation.^{4,35} We found a high frequency of spine deformity in our series (45.3%), coinciding with several reports,^{35–37} other series, however, have found higher rates.^{2,38,39} Spine deformity most often appeared in the frontal plane and least often in the sagittal plane. Acquired deformity (paralytic or idiopathic) was most common (42%), whereas congenital deformity comprised only 3% of the sample. There were few congenital spine deformities as compared to other series,^{40–43} probably because the patient groups studied were different.

The relationship between neurologic level involvement and presence of spine deformity was strongly significant: as neurological level descended, the prevalence of spine deformity decreased, a finding that this has been recorded in other series.^{41,42}

In 40.7% of patients pelvic obliquity was observed. Nine per cent of these showed no spine deformity at the last control, a fact that suggests a different aetiology for pelvic obliquity. Four per cent of patients were not able to maintain a stable sitting position and all patients with sitting instability were grouped together. We did not differentiate between patients who had lost free sitting function because of trunk imbalance secondary to spine deformity and those who were not able to sit because of severe brain lesions.

There was a clear relationship between hip joint status and neurologic level. A high percentage of patients with thoracic and high lumbar involvement presented hip dislocation. In thoracic level the cause has a postural basis, whereas in high lumbar level there is also a component of muscle imbalance. In our series the largest number of patients with hip dislocation had L3 involvement, the level that produces greatest hip muscle imbalance. In contrast, in Lindseth's work,⁴⁴ the greatest number of patients with hip dislocation had L1–L2 involvement. The differences between our series and Lindseth's are undoubtedly due to the fact that the attitude toward therapy for this pathology has taken more time to become unified in our setting. Our results cannot be compared with those of Sharrard's classic series⁴⁵ because the author applied a neurologic level classification different from those in general use. High neurological level patients showed hip flexion contractures of 25° or over more often. It is noteworthy that the patients who most frequently presented bilateral hip flexion contractures had thoracic and high lumbar levels. Once again, the cause in thoracic level patients is basically postural. The cause in lumbar level patients is double: first the important hip muscle imbalance, with strong flexor-adductor muscles and absence or weakness of the abductor-extensor muscles and second, postural factors, since the majority of these patients are not ambulatory and are seated the greater part of the day.

Many surgical procedures have been developed to improve hip function and radiological appearance.^{46–52} In the 1960's and 1970's reduction of all dislocated hips was standard, and children were submitted to several surgical interventions and long hospital stays.^{3,48} Failures were frequent, with repeated dislocation and joint stiffness after surgery.^{47,52} In the last decade, the indications for surgery in MMC have been reduced. In our series, surgery of the soft tissues is currently the most frequently used technique to treat fixed flexion contractures. It is accepted that bilateral luxation does not affect ambulation and type of orthosis, and therefore, does not have to be treated surgically.⁵¹ Treatment of orthopaedic deformities of the hips in patients with thoracic or high lumbar levels should be directed toward prevention and treatment of the contractures. Asher and Olson⁵¹ have demonstrated that at the L3–L4 level, contracture (not subluxation or dislocation) is the most significant factor affecting

the ability to walk. Surgery is usually reserved for patients with low lumbar or sacral level and unilateral dislocation, who are in good health, have normal IQs and are community ambulators.

Median age of walking onset in our series was earlier than in the only multicentre study published in our country.⁵³ Because of the high energetic cost of walking for thoracic, high lumbar and the great majority of mid-lumbar patients, many lose their ability to walk and became wheelchair-bound during the second decade of life. The median age at which patients lost the ability to walk in our sample was 128 months (10 years and 8 months), and is similar to other series. Shurtleff has introduced the concept of effective and independent mobility. This concept is defined as any efficient and effective means of moving about in space, and includes efficient upright ambulation as well as use of a wheelchair.² In our series this was accomplished at 43.3 months. As could be expected, patients with higher neurological levels began ambulation later and stopped earlier, and none of the low lumbar or sacral level patients ceased independent ambulation.

With regard to ambulatory status, 75% of patients were able to walk. The most significant factor for walking ability was the neurological level. In our series, 37.5% of patients with high or mid-lumbar levels were community walkers. This percentage is higher than those reported by DeSouza and Carroll (10%),⁵⁴ Hoffer *et al* (31%),¹⁴ Feiwell *et al* (26%),⁴⁸ Asher and Olson (25%),⁵¹ Stilwell and Menelaus (30%)⁵⁵ and Samuelsson and Skoog (23%).⁵⁶ Only Mazur *et al*⁵⁷ and Shurtleff *et al*² describe larger percentages of community ambulators (54 and 46.4%, respectively) in high or mid-lumbar patients. We conclude that neurological level is the main factor that determines neurological alterations, major orthopaedic deformities and ambulatory status.

References

- 1 Cuxart A. Situación actual de los pacientes adultos afectados de secuelas de malformación del tubo neural. Aspectos de integración personal y sociolaboral. Tesis Doctoral. Publicacions de la Universitat Autònoma de Barcelona. Bellaterra 1992.
- 2 Shurtleff DB. Myelodysplasias and exstrophies: significance, prevention and treatment. Grune & Stratton, Orlando, 1986.
- 3 Sharrard WJW. The orthopaedic management of spina bifida. *Acta Orthop Scand* 1975; **46**: 375–363.
- 4 Müller EB, Nordwall A. Prevalence of scoliosis in children with myelomeningocele in western Sweden. *Swine* 1992; **9**: 1097–1102.
- 5 Menelaus MB. *The orthopaedic management of spina bifida cystica*. 2nd ed. Edinburgh: Churchill Livingstone, 1980.
- 6 Iborra J. Deformidades del raquis en el mielomeningocele. Estudio de prevalencia y de historia natural. Tesis Doctoral. Publicacions de la Universitat Autònoma de Barcelona. Bellaterra 1995.
- 7 National Institutes of Health Consensus Development Panel on The Health Implications of Obesity. *Ann Intern Med* 1985; **103**: 147–151.
- 8 Roche AF, Siervogel RM, Chumlea WC. Grading body fatness from limited anthropometric data. *Am J Clin Nutr* 1981; **34**: 2831–2838.

- 9 MRC Grading Memorandum HMSO. 1976; **45**.
- 10 Weschler D. *The Weschler intelligence test*: revised manual. New York: Psychological corporation, 1983.
- 11 The terminology committee of the Scoliosis Research Society. A glossary of scoliosis terms. *Spine* 1976; **1**: 57–58.
- 12 Reimer J. The stability of the hip in children: a radiological study of the results of muscle surgery in cerebral palsy. *Acta Orthop Scand* 1980; (Suppl) 184.
- 13 Keggi JM, Banta JV, Walton C. The myelodysplastic hip and scoliosis. *Dev Med Child Neurol* 1992; **34**: 240–246.
- 14 Hoffer MM et al. Functional ambulation in patients with mielomeningocele. *J Bone Joint Surg* 1973; **55(A)**: 137–148.
- 15 Demografia i població. *Anuari Estadistic Barcelona* 1993, pp 38–39.
- 16 MRC Vitamin Study Research Group. Prevention of neural tube defects: results of the Medical Research Council Vitamin Study. *Lancet* 1991; **338**: 131–137.
- 17 Elwood JM, Elwood JH. *Epidemiology of anencephalus and spina bifida*. New York, Oxford University Press, 1980; pp 225–235.
- 18 Rickwood AMK. Congenital lesions of the spinal cord—General considerations. Neuromuscular problems in Orthopaedics. Ed. Galasko C.S.B. Oxford: Blackwell Scientific Publications, 1987.
- 19 Stein SC, Feldman JC, Friedlander M, Klein RJ. Is myelomeningocele a disappearing disease? *Pediatrics* 1982; **69**: 511–514.
- 20 Samuelsson L, Eklöf O. Scoliosis in mielomeningocele. *Acta Orthop Scandinavica*. 1988; **59**: 122–127.
- 21 Greene WB, Terry RC, DeMasi RA, Herrington RT. Effect of race and gender on neurological level in myelomeningocele. *Dev Med Child Neurol* 1991; **33**: 110–117.
- 22 Hall JG et al. Clinical genetic and epidemiological factors in neural tube defects. *Am J Hum Genet* 1988; **43**: 827–837.
- 23 Hayes JT, Gross HP. Orthopaedic implications of myelodysplasia. *J Am Med Association* 1963; **184**: 762–777.
- 24 Janerich DT. Female excess in anencephaly and spina bifida: Possible gestational influences. *Am J Epidemiol* 1975; **101**: 70–76.
- 25 Puente M et al. Estudi de creixement de la població infantil i adolescent de Catalunya (1986–1987). *Salut Catalunya* 1994; **8**: 121–128.
- 26 Dietz WH. Childhood obesity: susceptibility, cause and management. *J Pediatr* 1983; **103**: 676–686.
- 27 Hayes-Allen MC, Tring FL. Obesity: Another hazard for spina bifida children. *Br J Prev Soc Med* 1972; **27**: 192–196.
- 28 Manenica K. Identification of factors related to obesity in children with myelomeningocele. Master Thesis, University of Washington, 1982.
- 29 Sahuquillo J et al. Olygosymtomatic intracranial hypertension in patients with spina bifida and diagnostic of clinically arrested hydrocephalus. Results of a prospective study. *Eur J Paediatr Surg* 1993; **3** (Suppl I): 38.
- 30 Tew BJ, Laurence KM. The effects of admission to hospital and surgery on children with spina bifida. *Dev Med Child Neurol* 1976; **18** (Suppl 37): 18.
- 31 Anderson EM, Spain B. *The child with spina bifida*. London: Methuen, 1977.
- 32 Soare PL, Raimondi AJ. Intellectual and perceptual motor characteristics of treated myelomeningocele children. *Am J Dis Child* 1977; **131**: 199–204.
- 33 McLone DG, Czyzewski D, Raimondi AJ, Sommers RC. Central nervous system infections as a limiting factor in the intelligence of children with myelomeningocele. *Pediatrics* 1982; **70**: 338–342.
- 34 Azimullah PC, Smit LM, Rietveld-Knol E, Valk J. Malformations of spinal cord in 53 patients with spina bifida studied by magnetic resonance imaging. *Childs Nerv Syst* 1991; **7**: 63–66.
- 35 Lindseth RE. Myelomeningocele spine. In: Weinstein SL: (ed). *The pediatric spine*. Principles and practice. New York: Raven Press 1994; pp 1043–1068.
- 36 Schafer MF, Dias LS. Myelomeningocele: Orthopaedic Treatment. Baltimore, Williams and Wilkins, 1983; pp 12–14.
- 37 Shurtleff D, Goiney R, Gordon LH, Livermore N. Myelodysplasia: the natural history of Kyphosis and scoliosis. A preliminary report. *Dev Med Child Neurol* 1976; **18** (Suppl 37): 126–133.
- 38 Kahanovitz N, Duncal J. The role of scoliosis and pelvic obliquity on functional disability in myelomeningocele. *Spine* 1981; **6**: 494–497.
- 39 Raycroft JE, Curtis BH. Spinal curvature in myelomeningocele: Natural history and etiology. In: *The American Academy of Orthopaedic Surgeons. Symposium on Myelomeningocele*. St Louis: C.V. Mosby, 1972.
- 40 Mackel JC, Lindseth RE. Scoliosis in myelodysplasia. *J Bone Joint Surg* 1975; **57(A)**: 1031–1038.
- 41 Piggott H. The natural history of scoliosis in myelodysplasia. *J Bone Joint surg* 1980; **62(B)**: 54–58.
- 42 Banta JV, Bonani C, Prebluda J. The natural history of the scoliosis in myelomeningocele. Abstract. *Orthop Trans* 1986; **10**: 18.
- 43 Lonstein JE, Renshaw TS. Neuromuscular spine deformities. In: *Instructional course lecture*. American Academy of Orthopaedic Surgeons. St. Louis: CV Mosby, 1987.
- 44 Lindseth RE. Myelomeningocele. In: Morrisy RT (ed.) *Lovell and Winter's Pediatric Orthopaedics*, edn 3, Philadelphia: JB Lippincott 1990.
- 45 Sharrard WJW. Posterior iliopsoas transplantation in the treatment of paralytic dislocation of the hip. *J Bone Joint Surg* 1964; **46(B)**: 426–444.
- 46 Carroll NC. Hip instability in children with myelomeningocele. *Orthop Clin N Am* 1978; **9**: 403–408.
- 47 Feiwell E. Surgery of the hip in myelomeningocele as related to adult goals. *Clin Orthop Related Res* 1980; **148**: 87–93.
- 48 Feiwell E, Sakai D, Blatt T. The effect of the hip reduction on function in patients with myelomeningocele. *J Bone Joint Surg* 1978; **60(A)**: 169–173.
- 49 Kupka J, Geddes N, Carroll NC. Comprehensive management in the child with spina bifida. *Orthop Clin N Am* 1978; **9**: 97–113.
- 50 Carroll NC. Myelodysplasia. In: *Comprehensive Review Course for Orthopaedic Surgeons*. American Academy of Orthopaedic Surgeons. 58th Annual Meeting, April 2–7, 1989.
- 51 Asher M, Olson J. Factors affecting the ambulatory status of patients with spina bifida cystica. *J Bone Joint Surg* 1983; **65(A)**: 350–356.
- 52 Menelaus MB. Progress in the management of the paralytic hip in myelomeningocele. *Orthop Clin North America* 1980; **11**: 17–30.
- 53 Díaz I et al. Ambulation in patients with myelomeningocele: a study of 1500 patients. *Paraplegia* 1993; **31**: 28–32.
- 54 DeSouza LJ, Carroll N. Ambulation of the braced myelomeningocele patient. *J Bone Joint Surg* 1976; **58(A)**: 1112–1119.
- 55 Steiwell A, Menelaus MB. Walking ability in mature patients with spina bifida. *J Pediatr Orthop* 1983; **3**: 184–190.
- 56 Samuelsson L, Skoog M. Ambulation in patients with myelomeningocele: a multivariate statistical analysis. *J Pediatr Orthop* 1988; **8**: 569–575.
- 57 Mazur JM, Shurtleff D, Menelaus MB, Colliver J. Orthopaedic management of high level spina bifida. Early walking compared with early use in a wheelchair. *J Bone Joint Surg* 1989; **71(A)**: 56–61.