Mobility, Sitting Posture and Reaching Movements in Children with Myelomeningocele

BY

SIMONE NORRLIN
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Abstract

Children with myelomeningocele (MMC) usually have problems with daily life activities, but the background to their problems is not altogether obvious. An understanding of the possible causes of activity problems is a prerequisite for the effectiveness of physiotherapy. The overall aim of the present studies was to identify impairments above the celc level, which might influence mobility in children with MMC (study I) and to analyse sitting posture (study II) and the movement characteristics of reaching movements (study III and IV).

In total, 41 children and young adults with MMC and without mental retardation were investigated. Study I comprised 32 children, 6-11 years. Mobility and the caregiver assistance required for mobility were quantified according to the Paediatric Evaluation of Disability Inventory (PEDI) and correlation between mobility and neurological impairment, hand function and cognitive function were calculated. The results showed that nine children achieved independent mobility and that there was a moderate and significant correlation between the need for physical assistance and high celc level, impaired hand function and impaired cognitive function. In those children who used a wheelchair, only poor hand strength was significantly correlated with the need for caregiver assistance.

Study II comprised 11 children, 10-13 years, and a control group of 20 healthy children. Sitting posture was investigated by using a force plate and analysed from the frequency and the amplitude of the postural sway. The reaction forces before and during rapid arm lift were also analysed. The result showed that children with MMC had significantly lower sway frequency compared to the controls. In both groups, the ground reaction forces were registered before the children lifted their arms. Study III and IV comprised 31 children and young adults, 9-19 years and 31 matched controls. Reaching movements were investigated with a digitising tablet, linked to a computer. The ability to program and execute reaching movements was analysed and also the ability to adapt reaching to new visuomotor conditions. The results showed that the MMC group had poorer precision, less straight movements and shorter deceleration phases as compared to the controls. In both groups the movements were pre-programmed. In addition, adaptation of reaching to new visuomotor conditions was poor in the MMC group as compared to the controls.

In conclusion we found that impairments above the celc level influenced mobility and the control of sitting posture in children with MMC. Reduced precision and co-ordination of reaching, and also difficulties with motor adaptation, could partly explain their problems with hand activities. These findings need to be considered in therapy programs for children and young adults with MMC.

Keywords: myelomeningocele, PEDE, mobility, sitting posture, reaching, motor adaptation

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To my family
Anna, Christina, Louise and Peder
List of Papers

This thesis is based on the following papers, which will be referred to in the text by their Roman numerals.


III. Norrlin, S., Dahl, M., Rösblad, B. Control of reaching movements in children and young adults with myelomeningocele. *Dev Med Child Neurol* (accepted)

IV. Norrlin, S., Rösblad, B. Adaptation to a new visuomotor condition for reaching movements in children and young adults with myelomeningocele. (submitted)

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Abbreviations

MMC  myelomeningocele
TCS  tethered cord syndrome
ICIDH  International Classification of Impairment, Disability and Handicap
PEDI  Paediatric Evaluation of Disability Inventory
CoM  centre of mass
GRF  ground reaction force
AD  Alzheimer's disease
PD  Parkinson's disease
ADHD  Attention Deficit Hyperactivity Disorder
BMI  body mass index
TOMI  Test of Motor Impairment
VIQ  verbal IQ
PIQ  performance IQ
SD  standard deviation
EO  eyes open
EC  eyes closed
Hz  hertz
FB  feedback
NoFB  no feedback
WM  working memory
Introduction

Children with myelomeningocele (MMC) constitute a diverse group with varying manifestations of impairments. The most typical symptoms are the impaired sensorymotor function in the legs and reduced walking ability. However, it is well established that symptoms of impairments above the cele level are also common in these children, due to neuronal dysfunction and various malformations of the upper end of the central nervous system (11, 53, 66, 68, 83). Such symptoms are poor hand function (1, 35, 48, 49, 53, 68, 79), visuospatial dysfunction (19, 34, 78, 84) and cognitive dysfunction (19, 49, 78, 84).

In clinical practice it is evident that children with MMC, as a group, have problems with daily life activities, but the background to their problems is not altogether obvious. For example, children with the same cele levels might achieve different degrees of walking ability, which implies a multifactorial background to mobility problems. Or, children with the same cele levels might have a different sitting posture and sense of balance, which could be due to balance disturbances, not associated with the cele level. Furthermore, these children have been found to have clumsy and slow motor performance in hand activities, but the underlying movement characteristics of poor hand function have not been explored. The lack of an adequate explanation to all these problems, although well recognised among physiotherapists, implies that there are no uniform guidelines concerning their treatment in therapy programs. An understanding of the possible causes of activity problems is a prerequisite for the effectiveness of physiotherapy. These studies focus on impairments above the cele level which might influence mobility, sitting posture, and hand activities.

Myelomeningocele and associated structural abnormalities

MMC is a congenital malformation arising from failure of canalisation of the neural tube within the first month of pregnancy (68, 74). In Sweden, the incidence rate was 3.89 per 10 000 births in 1973-84, 2.89 per 10 000 births
in 1985-98 and 1.84 per 10,000 births in 1999-2000, showing that the incidence of MMC has declined over the last few decades (54). Today this means that about 20 children with MMC are born in Sweden each year. The malformation involves an open defect of the spine, where the spinal cord in its membrane sac protrudes as a cele from the back. The spinal defect may occur anywhere along the spinal column but it is usually located in the lower thoracic, the lumbar, or the sacral region of the spine (4, 68). The consequence of the spinal defect involve impaired motor and sensory function in the legs in varying degree, depending on the location and the size of the defect. As a rule it also entails a dysfunction of bladder and bowel control (4, 68). In addition, most children with MMC also have other anomalies in the central nervous system, such as Chiari II malformation.

Chiari II malformation and hydrocephalus

Chiari II malformation implies structural abnormalities of the brainstem and the cerebellum, including a small posterior fossa and a downward displacement of the lower brainstem, the cerebellum and the fourth ventricle through the foramen magnum (66, 68, 83). As a consequence, approximately 20-30% of the children with MMC develop severe symptoms of progressive brainstem dysfunction during the first three months of life, such as feeding and respiratory problems (11). In addition, those children have been found to have poor arm/hand function and problems with postural control and gross motor development (11).

Furthermore, Chiari’s malformation usually influences the circulation of the cerebrospinal fluid by blockage of the flow at the aqueduct or at the outlets of the fourth ventricle (4, 68, 83). This leads to the development of hydrocephalus, requiring treatment with a ventriculo-peritoneal shunt in about 80% of children with MMC (68). In the literature, the impairments associated with hydrocephalus and MMC are visuospatial dysfunction, oculomotor dysfunction, low performance IQ, and impaired eye-hand coordination (9, 19, 34, 44, 68, 77, 78, 84). Learning disabilities associated with shunt treated hydrocephalus have also been found in subjects with MMC (13, 59, 88).

Tethered cord syndrome and syringohydromyelia

All children with MMC have a tethering of the spinal cord to scar tissue and bony deformities, leading to stretching of the cord and its blood vessels (89). Some of the children develop tethered cord syndrome (TCS), most commonly apparent about the age of 6-12 years (68). The TCS is clinically
manifested by progressive neurological impairments such as muscle weakness, spasticity, changes in bladder and bowel control, sensory deficit, low back pain and also progressive musculoskeletal deformities (8, 11, 89).

About 20% of the children and young adults with MMC develop syringohydromyelia, implying cystic dilatation in the central canal of the spinal cord (8). Symptoms of syringohydromyelia are rare during infancy but develop gradually with increasing age. In children with MMC and scoliosis, examined by MRI, syringohydromyelia has been found to be present in about 40% of the subjects (66). The presence of syringohydromyelia has been found to be a risk factor for developing increasing symptoms of neurological dysfunction, such as progressive scoliosis, sensory deficits, muscle weakness or spasticity (8).

**Mobility**

Mobility has been defined as the individual’s ability to move about effectively in his surroundings, without the assistance of other individuals but, where appropriate, in a wheelchair or with other physical aids (ICIDH 1980, 86). This implies that mobility encompasses both the ability to transport the body in the surroundings and also the ability to change body positions. Mobility in children can be assessed with the PEDI, Paediatric Evaluation of Disability Inventory (28), a relatively new instrument which reflects a functional perspective on mobility, including moving about in a wheelchair.

The majority of children with sacral cee levels can walk, and children who can walk only have minor problems with mobility. However, children with lumbar or thoracic cee levels have reduced walking ability, and they need walking aids or a wheelchair. The presence of hydrocephalus and neurological abnormalities in the arms has been found significantly associated with reduced walking ability in children with MMC (80). Also, the presence of severe scoliosis has been found to be closely related to the inability to walk (66). Furthermore, in a previous study it was found that balance disturbances, spasticity in the legs and number of shunt revisions were significantly different between groups of children who achieved the expected walking ability and those who did not achieve expected walking ability (5).

The spinal defect in MMC is associated with reduced walking ability (3, 5, 32, 66), which might make a person more or less dependent on other
individuals in daily activities. However, the dependence of physical assistance required for mobility, as defined above, might also be associated with impairments above the cele level. The influence of hand function and cognitive function on mobility, whether you can walk or not, have not been clearly elucidated. Furthermore, only a few studies have investigated mobility, as defined above, in children with MMC.

Posture control

The control of posture involves the ability to orient body parts relative to one another, to the environment and to gravity (37, 73). Posture control with respect to gravity is essential in maintaining postural equilibrium, which has been defined as the state in which all forces acting on the body are balanced (37). Normally, postural equilibrium implies that the body posture is maintained in an intended position under static conditions and also during intended movements under dynamic conditions. Under upright and static conditions, the posture is characterised by small amounts of spontaneous sway, and postural sway can be used for evaluation of the vertical orientation with respect to gravity (73). Under dynamic conditions, a displacement of the centre of mass (CoM) usually tends to disturb the postural equilibrium. As a consequence, to compensate for the potential forthcoming CoM displacement that voluntary movement may cause, the movements are anticipated by postural adjustments (37, 43, 46, 73). For example, the task of reaching out to grasp an object can influence the body equilibrium, unless compensatory action is initiated before the arm is extended (37).

Organisation of posture control

Peripheral inputs from visual, somatosensory and vestibular systems register the body's orientation and movement in space with respect to gravity and to the environment (37, 43, 73). Visual inputs provide a reference for verticality, but are not absolutely necessary for posture control. The somatosensory system provides information about the position and motion of the body with respect to the supporting surface. The vestibular system, including peripheral receptors and vestibular nuclei in the brainstem, provides signals to the central nervous system related to the relative orientation of the head with respect to gravity. The vestibular system is assumed to play a major role in the control of posture (37, 43, 73).

Several authors have investigated the postural sway by analysis of the ground reaction forces (GRF) in subjects standing on a force plate (50, 60,
The sway can be quantified by both the amplitude and the frequency. The sway amplitude in children standing upright has been reported to decrease with age (61, 63, 85) and also to be smaller in standing with eyes open compared to standing with eyes closed. The frequency distribution of postural sway has also been reported to be age-dependent (61) and analysis of the sway frequency has been used to assess sensormotor development and to identify neural dysfunction in children (60). In a previous study, the sitting posture was investigated at six years of age in 15 children with MMC and a control group (38). The results showed that the children with MMC had significantly lower sway frequency as compared to the control group.

The properties of anticipatory adjustments have been illustrated by analysis of the GRF in both children and adults, while standing on a force plate and performing rapid arm lift (2, 7, 18, 62). The majority of the subjects demonstrated anticipatory changes of the GRF, occurring before the voluntary arm movement. Also, postural adjustments of reaching in sitting healthy adults have been investigated by Moore et al (52). They found that increased reach distance and decreased support were associated with earlier and larger postural adjustments. However, the authors concluded that there was no clear evidence that postural adjustments were required in advance in seated subjects performing arm lifts.

The body posture in children with MMC is deviant from that of non-disabled children, due to the muscle weakness in the legs. This is evident under both standing and sitting conditions. However, also other anomalies in the central nervous system might influence the ability to control the body posture. Whether children with MMC have a dysfunction in the postural control system above the cele level has not yet been shown.

Reaching movements

One important aspect of hand function is reaching toward a target, where the goal is to transport the hand with precision in both time and space. When reaching toward a target, the hand will describe a trajectory, which constitutes the movement path and the velocity profile of the hand. The path is normally a straight line from movement onset to the final part of the movement. The velocity profile is bell-shaped, and the peak velocity is scaled proportionately to target distance. If visual feedback is not provided during the course of the movement, the typical velocity profile can still be observed. These characteristics of the trajectory indicate that movement
parameters are mostly pre-programmed (21, 24, 27, 36). Prior to reaching, the internal representation of the movement is updated by visual information about the target and the hand. While visual feedback is not crucial during the course of the movement, it is of major importance for movement programming and for endpoint precision (21, 24, 27, 36). Several authors have suggested that there are discontinuities in the use of vision under reaching movements during development, up until around the age of eight years (16, 31, 65).

Organisation of reaching movements
The co-ordination of reaching is dependent on an intimate linkage between perception and action (6, 27, 33). The visual information about target location and upper limb position is used to specify the parameters (direction and extent) of the intended movement (20, 26, 41, 58, 64) and the somatosensory information about body parts and the environment is transformed and integrated into motor programs (40). The sensory transformations involve unique cortical and subcortical inputs to the motor cortex and the premotor areas — from the somatosensory cortex, the posterior parietal area, the basal ganglia, the cerebellum and the prefrontal cortex (40, 42, 69). The cerebellum is involved in the timing and co-ordination of hand movements and also participates in motor adaptation (23, 57, 71). In addition, the brainstem pathways are assumed to facilitate spinal motor neurons that innervate axial muscles, and brainstem neurons can also produce patterned and stereotyped movements (40, 70).

The ability to program and execute reaching movements in patients with Alzheimer's disease (AD) and Parkinson's disease (PD) has earlier been investigated by Ghilardi et al (24, 25). Their results indicated that visual information was used differently for movement control in the two groups of patients. Patients with AD lacked the ability to use feedforward commands and had to rely on ongoing visual feedback. Patients with PD, on the other hand, were disturbed by continuous visual feedback, which seemed to interfere with their movement execution. Yet another example is the study by Smyth et al (75) in which reach to grasp movements were investigated in children with developmental co-ordination disorder (DCD). The results showed that children with DCD had shorter movement time compared to controls, but they also had relatively shorter deceleration phases and higher movement velocity before target contact.
Adaptation of reaching

The ability to adapt to different visuomotor stimuli is crucial to effective and accurate movements (40). When movements under- or overshoot a target, for example in goal-directed reaching, the nervous system modifies the programming of subsequent movements to prevent errors (40, 57, 69, 71). This is called motor adaptation and involves changes in the control of movements that occur as a result of practice (10, 40). Voluntary movements are highly adaptable and improve in speed and accuracy with repeated trials of practice (40, 51, 57). When there are changes in the visuomotor conditions, movement error initially occurs, but after only a few trials the central nervous system adapts to the new condition, in order to minimise movement error.

Organisation of motor adaptation

Motor adaptation of reaching mainly involves the cerebellum and cerebellar cortical circuits, which participate in motor learning. The climbing fibres of the cerebellum are thought to register differences between expected and actual sensorymotor inputs, and when error in one movement is registered by the cerebellum, the motor program is adapted to the next movement (23).

The adaptation of reaching movements to changes in gain (extent) conditions has been investigated in healthy adults, by using a horizontal digitising tablet linked to a computer (58). Alterations in gain conditions produced the expected initial extent biases in the first trial. However, adaptation to the new gain occurred rapidly thereafter. Colvin et al (10) have recently examined motor adaptation of hand movements in children with MMC compared to children with attention deficit hyperactivity disorder (ADHD) and a group of healthy siblings. Contrary to the authors’ expectations, neither children with MMC nor those with ADHD displayed deficits in motor adaptation compared to their healthy siblings.

It is well established that many children with MMC have poor hand function, such as weak hand strength, impaired co-ordination and impaired sensory function in the hands (1, 29, 35, 48, 49, 53, 67, 78, 79, 80). Several authors found a correlation between MMC with hydrocephalus and poor hand function (67, 78, 79, 80). However, it has also been found that poor hand function in children with MMC was due to causes other than hydrocephalus (35, 53). Furthermore, Turner (79) found that poor hand function was not associated with the level of the lesion, while Mazur et al
(49) found that children with high cele levels had the most severely impaired hand function.

In studies of hand function researchers have used standardised functional tests, representing daily life activities like writing, cutting, unscrewing a jar, stacking coins or simulating eating. The outcome of these tests was based on the time taken for each task, and the main results showed that children with MMC had slow performance of hand activities. However, the slow motor performance could be due to several causes in children with MMC. The underlying movement characteristics, which might explain the slow performance, have not yet been investigated in goal-directed hand movements in children with MMC.
Aims of the studies

The aims of the present studies were to identify impairments above the cele level, which might influence mobility in children with MMC and to analyse sitting posture and also the movement characteristics of reaching movements. The specific objectives were:

- to investigate neurological impairment, hand function and cognitive function in a group of children with MMC and to identify factors of significance for independent mobility and for the physical assistance required for mobility in daily activities (study I)

- to investigate the horizontal GRF of seated postural sway and to analyse the GRF prior to and during rapid arm lift in children with MMC and a control group (study II)

- to investigate the ability to program and execute reaching movements in children and young adults with MMC and a control group (study III)

- to investigate the ability to adapt the movement extent of reaching movements to new visuomotor gain conditions in children and young adults with MMC and a control group (study IV)
Subjects and Methods

The majority of the children with MMC has long been regularly admitted to the Folke Bernadottehemmet for urological and medical investigation, from infancy into adulthood. Folke Bernadottehemmet is the regional child rehabilitation unit of Uppsala University Children’s Hospital. The referral area constitutes four counties and has about 1.2 million inhabitants. This means that about 80 children and young adults with MMC are admitted to the Folke Bernadottehemmet every year. In the last decade, a great deal of interest has been focused on functioning and disability associated with MMC. Consequently, multidisciplinary assessments by experienced professionals are routinely performed in daily clinical work.

In these studies, the investigations were performed during the subject’s referral period at the Folke Bernadottehemmet during the different study time periods. In four cases of study III and IV the investigations were performed at the local child rehabilitation unit. For inclusion in a study the subjects had been referred to the Uppsala University Children's Hospital for surgical closure of the celé and they were living in the regional area of the University Hospital (study I, III, IV) or in the county of Uppsala (study II). Subjects with mental retardation (IQ < 70) were excluded from the studies, since the assessments required maximal co-operation from the subjects. Nine subjects participated in all four studies. The number of participants, age, gender and the overlap in study groups are shown in Table 1.
Table 1. Number of participants, age, gender and overlap in study groups

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<td>9 study II</td>
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Factors of significance for mobility in children with myelomeningocele (study I)

Study I was part of a larger project where all children with MMC, aged 6-11 years and living in the region during the time period October 1995 to October 1997, were invited to participate in a multidisciplinary study. Forty children fulfilled the inclusion criteria and were invited to participate. Nine families failed to attend for practical or personal reasons. In addition, one out-region child was invited and agreed to participate. Thus the study comprised 32 children, 19 boys and 13 girls (median age 8.7 years). The characteristics of the total samples of study I-IV are shown in Table 2.

Data collection

The data were collected during a two-week period, when the child was staying with one or both parents at the Folke Bernadottehemmet. Mobility was assessed with the Paediatric Evaluation of Disability Inventory (PEDI, 28) and scored as caregiver assistance. The instrument was designed for evaluation of functional skills and caregiver assistance in children, from six months to 7.5 years of age. However, it can be used for evaluation in older children if their performance falls below that expected of non-disabled children (28). Functional skills measure the capacity to perform activities of daily life in the child’s environment and caregiver assistance measures independence or the amount of physical assistance required of the caregiver in order for the child to accomplish these activities. The performance of each item was quantified on a six-point scale (5-0) and classified as: independent,
supervision, minimal assistance, moderate assistance, maximal assistance and total assistance. The item scores were summarised in raw scores and the raw scores were converted into scaled scores, using the PEDI software program (28). The scaled scores account for item difficulty and are not adjusted for age. Scaled scores describe the child’s performance relative to the maximum possible scores (0-100). The PEDI has been reported as reliable and valid for assessment of functional performance in children with disabilities (55). In a Swedish study, the results obtained from the PEDI were compared with American normative data, and it was suggested that the American normative data were appropriate for reference purposes in Sweden (56). The PEDI assessments were administered by the author.

Factors of importance for mobility were selected from clinical experience and from earlier studies. These factors were categorised into neurological impairment, hand function, and cognitive function. Information about early and severe brainstem dysfunction, shunt treated hydrocephalus, scoliosis, TCS and body mass index (BMI) was collected from the medical records. Brainstem dysfunction was said to be present if the child had had symptoms related to the brainstem level before three months of age, such as feeding and respiratory problems. TCS was manifested as progressive neurological and orthopaedic deterioration in the legs or changes in the urodynamic pattern together with a low-lying conus medullaris. Brainstem dysfunction and TCS were assessed by the neurologist.

Hand function was investigated with respect to muscle strength and co-ordination. Isometric hand strength was registered in a standardised situation with a Jamar dynamometer and expressed in % of normal values. Hand co-ordination was assessed in three items (one-hand speed, bimanual speed, one-hand precision) of the Test of Motor Impairment (TOMI, 76). Hand function was assessed by the author, who also assessed the functional cele level and walking ability. The cele level was defined as the lowest level on the better side at which the child was able to perform an antigravity movement through the available range of joint motion (45). Walking ability was defined according to Hoffer et al (32) as four categories: community, household, non-functional and non-ambulators. Cognitive function was investigated with respect to intelligence, executive function and visuospatial function. Verbal and performance intelligence quotients (VIQ, PIQ) were assessed with the Wechsler Intelligence Scale for Children (81, 82). Executive function (attention, control, organisation) was assessed with the Nepsy - part 1 and visuospatial function was assessed with the Nepsy - part 4 (39). The standard deviations (SD) of each item were summarised and the
mean score was calculated. Cognitive function was assessed by the neuropsychologist.

Table 2. The total samples of study I-IV

Study I included one child living outside the referral area, study II included only children living in the county of Uppsala, study III and IV included two children living outside the referral area and also one child who was not referred to the University Children's Hospital for surgical closure of the cele.

<table>
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<th>Study</th>
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<td>Non-responders</td>
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<td>7 (78%)</td>
<td>3 (33%)</td>
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<td>II</td>
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<td>12</td>
<td>11</td>
<td>9 (82%)</td>
<td>3 (27%)</td>
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<td>1</td>
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<tr>
<td>III &amp; IV</td>
<td>Total sample</td>
<td>52+3</td>
<td>28+3</td>
<td>21 (68%)</td>
<td>5 (16%)</td>
<td>6 (19%)</td>
</tr>
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<td></td>
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<td></td>
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<tr>
<td></td>
<td>Non-responders</td>
<td>24</td>
<td>18 (75%)</td>
<td>5 (20%)</td>
<td>6 (25%)</td>
<td>7 (29%)</td>
</tr>
</tbody>
</table>

Brainstem symptoms = early and severe symptoms of brainstem dysfunction
Cele level defined according to Lindseth (45)

Force measurements of postural sway and rapid arm lift in seated children with myelomeningocele (study II)

All the children with MMC, 10-13 years and living in the county of Uppsala during the time period August 1998 to March 1999, were invited to participate in study II. The total sample constituted 12 children (Table 2). One girl chose not to attend and thus the study comprised 11 children, nine boys and two girls (median age 11.4 years). Study II also comprised a control group of 20 healthy children without any known disabilities, 10 boys and 10 girls (9-10 years), who were recruited from a local school.
Data collection and analysis

The measurements took place at the University Hospital and were performed after careful information to the parent and child. For data collection we used the Vifor-system, consisting of a Kistler force plate synchronised with two video cameras and a personal computer. To enable force plate measurements in sitting, a chair was specially constructed to fit the force plate. Testing of the method preceded this study and is further described by Karlsson et al (38). The children were sitting on the chair with no back or foot support. They were instructed to sit upright and to look straight ahead at a visible target. The GRF of the postural sway was registered under three different conditions: sitting as still as possible with eyes open (EO), sitting as still as possible with eyes closed (EC), sitting with EO and lifting both arms to the shoulder level as fast as possible and to point at a target in front of them. The measurements were conducted by the author.

Data were collected during 30 s at a sampling frequency of 50 Hz. The horizontal GRF was analysed in the anteroposterior (x) and the mediolateral (y) directions. The GRF was normalised with the body mass and thus the forces were expressed as the corresponding acceleration of the CoM (mm/s²). The sway amplitude was quantified by the standard deviation (SD) of the centre of mass (CoM) acceleration, and the sway frequency was quantified by the oscillations (Hz) of the CoM. Under the arm lift condition the GRF was analysed one second (s) before and one s after the fastest arm lift of each child. The fastest lift and movement time (ms) were estimated from the video images, recorded in intervals of 20 ms. GRF onset (-ms) was defined when the CoM acceleration first exceeded 2 SD of the anteroposterior sway amplitude during quiet sitting in each subject. The GRF onset and the peak amplitude (m/s²) were measured relative to movement start and movement time.

Control of reaching movements in children and young adults with myelomeningocele (study III)

The total sample comprised all the children and young adults with MMC in the study population, who were living in the referral area during the time period May 2000 to January 2001 (Table 2). In addition, two subjects who were living outside the referral area and also one child who had not been referred to the University Children's Hospital for surgical closure of the cele, but were regularly referred to Folke Bernadottehemmet, were invited and agreed to participate in study III. Two children did not attend for practical
and personal reasons, and 22 subjects were not invited to participate, since they were not referred to the Folke Bernadottehemmet during the study time period. Thus the study sample comprised 31 subjects, 18 boys and 13 girls (mean age 12.9 years). Study III also comprised a control group of 31 healthy subjects, matched for age and gender, and recruited through contact with all categories of the personnel at the Folke Bernadottehemmet.

Data collection and analysis
The data were collected during the participant's regular stay at the Folke Bernadottehemmet. For data collection we used a horizontal digitising tablet (42x30 cm) and a hand-held digitizer (12x6x2 cm), linked to a computer with a screen size of 26x19 cm. The location of the digitizer on the tablet (x and y co-ordinates) was displayed on the computer screen as a cursor. Circles on the screen indicated the starting point and the target positions, located 2.4 cm, 4.8 cm and 7.2 cm from the starting point. The scaling factor (gain) of the tablet to the screen was 47%, which means that reaching the screen targets required hand movements of 5.1 cm, 10.4 cm and 15.3 cm, respectively.

The targets were presented on the screen one at a time in a pseudo-random sequence at 45 degrees from the midline to the contralateral side of the body. In the testing procedure the subjects made planar reaching movements towards the targets by sliding the digitizer along the surface of the tablet. The movements were registered under two conditions - first with visual feedback (FB) and second without visual feedback (NoFB). Each subject made 30 trials with FB and 30 trials with NoFB. Under the NoFB condition the screen cursor position was blanked after movement onset in order to preclude visually based corrections of the movement. At the end of each trial with NoFB, knowledge of result was provided by displaying the movement path on the screen. This method has been further described by Ghez et al (21). The measurements were conducted by the author.

Data were sampled at 200 Hz and the analysis was performed with a computer program developed by Gordon et al (27). Movement programming was analysed from the velocity profiles, and movement execution was analysed from the spatial and temporal parameters of the movement paths. Automatic computer routines were used to mark movement onset and endpoint. These critical points were checked manually, and a few incorrect trials were rejected. As a global measure of the spatial precision we used movement Error, divided into Extent error (cm) and Directional error (degree), which were analysed separately. Automatic computer routines
were also used to calculate a Linearity index and a Path length ratio and also the movement Duration (ms), the Peak velocity (cm/s) and the Time to peak velocity (ms). In addition, we calculated the time spent for deceleration, defined after the calculation of the relative proportion between Time to peak velocity and movement Duration (%).

Adaptation of reaching movements in children and young adults with myelomeningocele (study IV)

Study IV was carried out in connection with study III and comprised the same total sample and the same control group (Table 2). In study IV, the data of one child with MMC could not be interpreted into the analysis due to technical problems. This was one of the five children who had had early and severe symptoms of brainstem dysfunction. We then excluded the data from this child and also the data from the matched control child. This means that the analysis in study IV was based on data from 30 subjects with MMC and 30 subjects without disabilities.

Data collection and analysis

For data collection we used the same equipment as described above. In study IV, all the measurements were performed without visual feedback, which means that visual information about movement error was only available after each trial, when the movement path was displayed on the screen. The reaching movements were executed under two different gain conditions - a base condition and an altered condition. One session included 90 trials, first 30 trials under the base condition and then 45 trials under the altered condition, followed by 15 trials under the base condition again. Under the base condition (trial 1-30), the gain (scaling factor) of the path display was 0.47:1 (screen:hand) and under the altered condition (trial 31-75) the gain of the path display increased to 0.94:1. The increased gain was designed to cause an initial overshoot of movements and the return to the base condition (trials 76-90) would then imply an initial undershoot of movements. When the altered condition was introduced to the first subject, the computer program was interrupted because the overshoot was too large. Therefore, all the subjects were informed that there would be a gain change after trial 30. However, after the altered condition (31-75), the gain change was implemented unexpectedly.
Data were sampled at 200 Hz, and the analysis was performed with a computer program developed by Gordon et al (27). Automatic computer routines were used to mark movement onset and endpoint. These critical points were checked manually, and a few incorrect trials were rejected. The movement outcome was calculated from the extent error of each trial, computed as the difference between the actual movement extent and the distance to the target. In order to obtain comparable measures of extent error toward different targets, we normalised them with respect to the target distances, and error was expressed as percentage error of the target distance. To begin with, the percentage error was analysed from the base condition (trial 1-30) and then the measurements were divided into groups of trials, and different phases of the measurements were analysed.

Study population

The study population was defined as all children and young adults with MMC and without mental retardation, referred to the University Children's Hospital for surgical closure of the cele, living in the referral area and born between 1981-1991. To investigate if the samples were representative of the study population we compared the non-responders with all the subjects who had participated in at least one study. The two subjects who were living outside the referral area and also the child who had not been referred to the University Children's Hospital for surgical closure of the cele are not included in the study population. The characteristics of the study population, based on all four studies, is shown in Table 3. Fifty-two subjects fulfilled the inclusion criteria of the study population. Fourteen families chose not to attend for practical or personal reasons. Thus the study population comprised 38 subjects, 21 male and 17 female, who agreed to participate in different studies. Comparing the participants with the non-responders, there were some difference in age categories. The participants represented more young children and fewer young adults. The participants also involved more subjects with medium-high cele levels but fewer subjects with high and low cele levels as compared to the non-responders. Only minor differences were found for early and severe brainstem dysfunction or shunt treated hydrocephalus. Generally, the participants were representative of children and young adults with MMC in the referral area. The distribution of participants in the different studies is shown in Fig 1.
Table 3. The study population

<table>
<thead>
<tr>
<th></th>
<th>Participants</th>
<th>Non-responders</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age category 1</td>
<td>3 (8%)</td>
<td>5 (36%)</td>
<td>8 (15%)</td>
</tr>
<tr>
<td>Age category 2</td>
<td>18 (47%)</td>
<td>6 (43%)</td>
<td>24 (46%)</td>
</tr>
<tr>
<td>Age category 3</td>
<td>17 (45%)</td>
<td>3 (21%)</td>
<td>20 (38%)</td>
</tr>
<tr>
<td>Gender (male)</td>
<td>21 (55%)</td>
<td>7 (50%)</td>
<td>28 (54%)</td>
</tr>
<tr>
<td>Shunt</td>
<td>29 (76%)</td>
<td>10 (71%)</td>
<td>39 (75%)</td>
</tr>
<tr>
<td>Brainstem symptoms</td>
<td>7 (18%)</td>
<td>3 (21%)</td>
<td>10 (19%)</td>
</tr>
<tr>
<td>Th-L2</td>
<td>7 (18%)</td>
<td>5 (36%)</td>
<td>12 (23%)</td>
</tr>
<tr>
<td>L3-L4</td>
<td>16 (42%)</td>
<td>1 (7%)</td>
<td>17 (33%)</td>
</tr>
<tr>
<td>L5-S</td>
<td>15 (39%)</td>
<td>8 (57%)</td>
<td>23 (44%)</td>
</tr>
</tbody>
</table>

Age category 1 = born 1981-83
Age category 2 = born 1984-87
Age category 3 = born 1988-91
Brainstem symptoms = early and severe symptoms of brainstem dysfunction
Cele level defined according to Lindseth (45)

Inclusion criteria:
- Without mental retardation
- Surgical closure of the cele at the University Hospital
- Living in the referral area
- MMC born between 1981-1991

Fig 1. The study population and the distribution of participants in study I-IV
Statistical methods
Non parametric methods were used in Study I and II, with the Mann-Whitney U test for differences between two independent groups and the Spearman rank order correlation coefficient for associations between variables. Differences within the MMC group were calculated with the Kruskal-Wallis test in Study II. Descriptive statistics were presented as median values and ranges. In Study III and IV, analysis of variance (ANOVA) was employed for comparison of mean values for different trials and conditions. If sphericity of the error covariance was not assumed, the intra-subjects effects were based on the Greenhouse-Geisser adjustment for non-sphericity. Group differences were also analysed with $t$-tests and paired $t$-tests in study IV. The level of statistical significance was set at $p<0.05$. 
Summary of Results

Study I

Mobility was investigated in 32 children. Nine children scored independent mobility (100) and seven of them had sacral cele levels and were community ambulators. The independent children had significantly better hand function, visuospatial function and cognitive function compared to those who needed caregiver assistance. The differences in hand strength, VIQ and PIQ are shown in Fig 2. A moderate and significant correlation was found between the need for caregiver assistance, on the one hand, and high cele level, poor hand function and impaired cognitive function, on the other hand. The correlation coefficients between the caregiver assistance scores and the independent variables are shown in Table 4. The children with severe and early symptoms of brainstem dysfunction and the children with scoliosis needed more caregiver assistance compared to the others ($p<0.001$, $p=0.008$).

Table 4. The correlation coefficients between caregiver assistance scores and walking ability, cele level, hand function and cognitive function in 32 subjects

<table>
<thead>
<tr>
<th>Variables</th>
<th>$r_s$</th>
<th>p-values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Walking ability</td>
<td>0.656</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Cele level</td>
<td>0.639</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Hand co-ordination</td>
<td>0.593</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Hand strength</td>
<td>0.514</td>
<td>0.003</td>
</tr>
<tr>
<td>Visuospatial perc.</td>
<td>0.444</td>
<td>0.011</td>
</tr>
<tr>
<td>Executive function</td>
<td>0.415</td>
<td>0.018</td>
</tr>
<tr>
<td>Performance IQ</td>
<td>0.411</td>
<td>0.019</td>
</tr>
<tr>
<td>Age</td>
<td>0.218</td>
<td>0.231</td>
</tr>
<tr>
<td>Verbal IQ</td>
<td>0.134</td>
<td>0.464</td>
</tr>
<tr>
<td>Body Mass Index</td>
<td>0.100</td>
<td>0.587</td>
</tr>
</tbody>
</table>
To continue, the material was then analysed when the 11 children who were community ambulators were excluded, since mobility is usually less complex when you are able to walk. The new subgroup consisted of the 21 children who used a wheelchair and/or walking aids, categorised as household to non-ambulators. In this second group, only hand strength was significantly correlated with the caregiver assistance scores ($r_s=0.703$, $p<0.001$). However, the celc level was not significantly correlated with the caregiver assistance scores ($r_s=0.379$, $p=0.09$) and neither were any of the other selected variables. Furthermore, there was a tendency for the girls to need more caregiver assistance than the boys ($p=0.050$).

Fig 2. Hand strength (% of normal values), verbal IQ (VIQ) and performance IQ (PIQ). The black bars represent the nine independent children and the white bars represent those who needed caregiver assistance. The median values are shown above each bar.

Study II

Postural sway

The postural sway was investigated in 11 children with MMC and 20 children without disabilities. The MMC group had lower frequency of the anteroposterior GRF under both conditions EO ($p=0.0004$) and EC ($p=0.0015$), as compared to the control group. The distribution of the anteroposterior frequency is shown in Fig 3. The lowest anteroposterior frequencies with EO were found in the children with early brainstem dysfunction (n=3) and in the children with scoliosis ($p=0.044$). The frequency distribution within the MMC group was not significantly different.
regarding shunt treatment, and the frequency was not significantly correlated
with cele levels or BMI. When the cele levels were grouped into two
categories, low (S-L5) or medium-high to high (L4-Th), a significant
difference in sway frequency was only found in the anteroposterior direction
with EC (Fig 4). The sway amplitude was not significantly different between
the MMC group and the control group for either condition EO or EC.
However, the sway amplitude in children with medium-high and high cele
levels, as compared to those with low levels, was significantly larger except
for in the anteroposterior direction with EC (Fig 5). In addition, the sway
amplitude increased more with EC in the control group compared to the
MMC group, and the group difference was close to significant in the
mediolateral direction ($p=0.060$). Children with scoliosis had larger sway
with EO in both directions ($p=0.045$ and $p=0.011$). As for children with
early brainstem dysfunction or shunt treatment, the sway amplitude was not
larger and the sway amplitude was not significantly correlated with the BMI.

![Distribution of the sway frequencies in the anteroposterior direction with eyes open and eyes closed in the children with MMC (n=11) and the control group (n=20)](image)
Fig 4. The sway frequency within the MMC group, when the cele levels were categorised into low (S-L5) and medium-high to high (L4-Th). The error bars show 95% CI.

Fig 5. The sway amplitude within the MMC group, when the cele levels were categorised into low (S-L5) and medium-high to high (L4-Th). The error bars show 95% CI.
Rapid arm lift

The movement time of arm lift was longer for the children in the MMC group \((p=0.0001)\). The initial GRF was directed forward in all trials for the MMC children and in all but one for the controls. In both groups the onset of GRF occurred before the subjects lifted their arms. During arm lift the GRF was characterised by two peaks (Fig 6), a forward acceleration of the CoM and a backward CoM acceleration (deceleration) - occurring at about the time when the arms reached the horizontal plane.

![Figure 6. A typical CoM acceleration curve during arm lift. The vertical line represents the movement start and the initial positive acceleration of the CoM represents a forward direction of the GRF.](image)

Study III

Movement programming

Reaching movements were investigated in 31 subjects with MMC and 31 matched controls. In both groups the velocity profiles were bell-shaped and scaled proportionately to target distances under both visual conditions, indicating that reaching movements were pre-programmed \((F_{2,59} =298.1, p<0.001)\). No statistically significant interactions for Peak velocity were found between FB and group \((F_{1,60}=2.4, p=0.123)\) or between trial and group \((F_{2,59} =2.4, p=0.095)\).
Movement execution

The spatial precision was poor in the MMC group compared to the controls, indicated by larger movement Error under both visual conditions ($F_{1,60} =10.601, \ p=0.002$). There was also a main group effect for Linearity ($F_{1,60} =5.940, \ p=0.018$), showing that movements had larger deviations from a straight-line path in the MMC group compared to the control group. In addition, there was a main group effect for Path length ratio ($F_{1,60} =8.356, \ p=0.005$), showing that the movement paths were longer in the MMC group compared to the controls. Fig 7 presents the movement paths of one child in the MMC group and one in the control group.

![Movement paths](image)

Control   MMC

Fig 7. Movement paths from the start-point to the three targets under the visual feedback condition

No main effect of group was found for movement Duration ($F_{1,60} =0.007, \ p=0.935$). However, movement Duration decreased more in the MMC group in NoFB trials, indicated by a close to significant interaction between FB and group ($F_{1,60} =3.8, \ p=0.055$). In addition, the subjects with MMC had significantly longer Time to peak velocity relative to movement Duration, which means shorter deceleration phases ($F_{1,60} =8.906, \ p=0.004$). When looking at the velocity profiles of each subject it was obvious that the profiles were sometimes discontinuous in the MMC group, especially under the FB conditions. This is illustrated in Fig 8, which shows the velocity profiles under both visual conditions of one child with MMC.

![Velocity profiles](image)

Without FB   With FB

Fig 8. Velocity profiles under both conditions without and with visual feedback in one girl with MMC

25
Differences within the MMC group

Generally, subjects with symptoms of early and severe brainstem dysfunction (n=5) were slower and had poor precision compared to subjects without symptoms of brainstem dysfunction. However, a main group effect was found only for Time peak velocity/Duration ($F_{1,29} = 7.119$, $p=0.012$), showing that subjects with symptoms of early brainstem dysfunction had shorter deceleration phases under both visual conditions. Subjects who were shunt treated (n=21) were not generally slower compared to subjects who were not shunt treated. However, for movement Duration there was an interaction between shunt and FB ($F_{1,29} = 10.2$, $p=0.003$) and also for Time to peak velocity ($F_{1,29} = 9.0$, $p=0.005$), indicating that subjects with shunt were slower under the FB condition but faster under the NoFB condition, compared to subjects without shunt.

Study IV

Adaptation of reaching was analysed in 30 subjects with MMC and 30 matched controls. Under the base condition (trial 1-30), both groups produced relatively accurate movements, and the difference in percentage error between the groups was not significant ($p=0.346$). The changes in gain condition produced the expected initial extent biases in both groups as shown in Fig 9. The first change in gain (trial 31) produced an increase in extent, and the second change back to the base condition (trial 76) produced a decrease in movement extent. The mean percentage error in trial 31, after the subjects had been informed about the forthcoming gain change, was significantly larger in the MMC group as compared to the control group ($p=0.001$). The control group did not even reach the target in trial 31 (Fig 9). In trial 76, after the unexpected gain change, the percentage error was not significantly different between the groups ($p=0.250$).
Fig 9. Mean percentage error of trials toward targets at three different distances. The error bars show 95% CI. White squares represent the control group and black squares represent the MMC group.
Movement adaptation

Under the altered gain condition (trial 31-45) the percentage error was significantly larger in the MMC group compared to the control group ($F_{1,53}=8.547, p=0.005$). In the control group, movements were rapidly adapted to the altered condition (Fig 9). There was no significant difference in percentage error of trial 37-46 compared to the base condition (trial 22-30) in this group ($p=0.311$). However, in the MMC group the percentage error of trial 37-46 was significantly larger ($p<0.001$) compared to the base condition. Analysis of the final adaptation, as analysed from the last 15 trials under the altered condition (trial 61-75), showed that subjects in the MMC group could not fully adapt their movements to the new gain. Their mean percentage error continued to be larger, as compared to the base condition ($p<0.001$). In the control group, the final adaptation was illustrated by no difference in percentage error between the last 15 trials under the altered gain condition and the base condition ($p=0.152$).

Differences within the MMC group

Some differences were found when the MMC group was analysed with respect to cele level, shunt treatment and early and severe symptoms of brainstem dysfunction. Subjects with symptoms of brainstem dysfunction had larger percentage error compared to the others (Fig 10), but since the results of only four children with brainstem dysfunction were analysed, no statistical analysis of these measurements was performed. Subjects who were shunt treated did not have statistically significantly larger percentage error under the base condition ($p=0.601$) or under the altered condition ($p=0.434$) as compared to those without hydrocephalus (Fig 11).
Fig 10. Mean percentage error of 90 trials, merged into groups of five. The error bars show 95% CI. Black squares represent subjects with symptoms of early and severe brainstem dysfunction, grey squares represent subjects without brainstem dysfunction and white squares represent the control group.

Fig 11. Mean percentage error of 90 trials, merged into groups of five. The error bars show 95% CI. Black squares represent subjects with shunt treated hydrocephalus, grey squares represent subjects without hydrocephalus and white squares represent the control group.
Discussion

In the present studies we found that mobility in daily activities was associated with hand function and cognitive function, apart from the cele level and walking ability. It was obvious that children with poor hand strength who used a wheelchair also needed a lot of caregiver assistance in mobility. In addition, the control of sitting posture was characterised by a low sway frequency in children with MMC, implying slow motor performance. The results also showed that the subjects with MMC had problems with reaching movements, which could not be explained by impaired movement programming. Instead, this was related to the execution of on-going movements, most evident under conditions when visual feedback was provided. Furthermore, the subjects with MMC could not rapidly adapt reaching movements to a new visuomotor condition, as could the subjects without disabilities.

Methods

Subjects
The study population constituted a selected material, since only subjects who fulfilled the inclusion criteria were included. In addition, only subjects who were referred to the Folke Bernadottehemmet during the study time periods participated in the studies. This was for practical reasons, meaning that the investigations involved only minor efforts for the participants and their parents. Thus, the number of participants in the studies was relatively small (n=38), constituting 73% of the study population (n=52). Generally, the participants were representative of children and young adults with MMC in the referral area. Still, the small size of the study sample needs to be considered when interpreting the results. In addition, it is hard to draw general conclusions about the entire population of children and young adults with MMC, since there are several confounding factors to motor performance in daily activities. These could include differences in personal and environmental factors and differences in the medical and therapeutic
treatment in different parts of the country. Furthermore, the present study material is relatively different compared to older studies, since almost all the children with MMC survive today, even those with severe structural malformations of the central nervous system.

Statistical considerations
Despite the fact that the study samples were small, several significant differences were found between independent groups. However, there might have been type II errors when small groups were compared, implying that we failed to explore true differences. In study II and IV, when the groups were smaller than five subjects, the differences are presented in text or figures. To create larger groups we also categorised the cele levels into high, medium-high and low cele levels.

Data collection
For the data collection we used standardised instruments, and the assessments were performed by experienced professionals. For assessment of mobility we used the PEDI (study I), because it reflects a functional perspective on mobility, including both walking ability and moving about in a wheelchair. We found that the PEDI was an instrument well suited for this group of children. We also found that you must look at each item separately, not only the total scores, if you want to get a realistic picture of the amount of caregiver assistance required. For example, if one child only needs supervision in several items, he or she might get the same total score as another child who needs maximal assistance in one of the items. In this study we investigated the influence of neurological dysfunction, hand function and cognitive function on mobility. However, the impact of environmental and personal factors has not been considered, since mobility was investigated during the referral period at Folke Bernadottehemmet.

The method of analysing postural sway (study II) was tested in a previous study (38) which showed that frequency analysis could be used to detect differences between seated children with and without MMC. The clinical importance of sway measurements in assessing posture control in sitting has been questioned, since the base of support is larger than in standing. Yet, in this study the children were instructed to sit as still as possible during the measurements. Force measurement with the Vifor system was found uncomplicated and not time consuming for the children. Force plate
measurements are suggested as a useful method in combination with other clinical and laboratory tests for the purpose of evaluating sitting posture.

The digitising tablet used for kinematic analysis of reaching (study III and IV) has previously been used by several authors to investigate movement programming and execution (14, 15, 21, 24, 25, 27). By using this instrumentation we could explore some underlying movement characteristic in subjects with MMC. The tasks reminded subjects of computer games, and the method was well suited for and tolerated by the subjects. The investigations were easy to accomplish and not very time consuming. These two investigations were carried out in succession with only a short break in between. In study III, reaching movements were performed under the same gain condition as in the base condition of study IV, which means that the subject had had ample time of learning the visuomotor transformation from table to screen before the second investigation.

Mobility
Children with MMC are usually classified according to cele level, which does not provide complete information regarding mobility and motor functioning. Four children with low cele levels needed caregiver assistance in daily activities while two children with higher cele levels achieved independent mobility according to the PEDI. Furthermore, when the 21 children who used a wheelchair for mobility were analysed separately, only hand strength was significantly correlated with mobility. Hand strength ranged from 41% to 120% of normal values in this group. Reduced hand strength has previously been reported in children with MMC, compared to a population without disability (1, 53). However, the importance of hand strength for the caregiver assistance required in daily activities has not been reported. Hand strength might be of such importance that other factors such as cele level, hand co-ordination and cognitive function have less influence on mobility in children who use a wheelchair. This seems quite natural, and it also indicates that hand strength should be taken into account in mobility training.

The results of study I also showed that the children who had had severe and early symptoms of brainstem dysfunction (n=6) needed a lot of assistance, including one child with a sacral cele. These children were observed to have obvious low muscle tone, and their hand strength was highly impaired. We
have found no earlier studies investigating caregiver assistance in mobility, as defined in the PEDI. However, several authors have found poor hand coordination and cognitive function in children with MMC (9, 17, 19, 34, 53) and this was confirmed by the results of study I.

Posture

The results of study II showed significantly lower sway frequencies in children with MMC compared to children without disabilities. This is in accordance with the previous study of children at six years of age (38). The difference between groups was larger at 10 years than it was at six years. Low frequency of postural sway has also been found in children with visual impairment (60). However, the pathogenic mechanism of a low sway frequency has not been clearly elucidated. Two different modes of perceiving postural relations have been suggested, one stemming from the vestibular system and one from the somatosensory system (37). The vestibular system takes the head as reference and registers high-frequency components of movements, while the somatosensory system takes the stable ground as a reference and registers low-frequency movements. The cele and the interrupted spinal control mechanism imply a dysfunction of the somatosensory system. However, the sway frequency was not significantly correlated with the cele level. Instead, the children with symptoms of early brainstem dysfunction had the lowest sway frequencies, implying a dysfunction in the vestibular system. Furthermore, except for one boy with hemi-cele, all children in the MMC group had lower anteroposterior sway frequency, as compared to the median of the controls. This indicates that they might not be able to make fast high-frequency adjustments of posture.

The sway amplitude with closed eyes was smaller for several children in the MMC group and also for some of the controls. This is in contrast with earlier studies, showing a larger sway amplitude in standing subjects during conditions EC compared to EO (60, 61, 85). However, normal children, 4-6 years of age, have also been demonstrated to sway more with EO (60, 61). This difference was explained by an improvement in the use of vision as children grow older (10-12 years). One explanation could be that the higher sway amplitudes with EO in children with MMC were due to immature integration and processing of visual inputs.

The movement time of arm lift was significantly longer for the children in the MMC group compared to the controls. This can be due to the low force
of the shoulder muscles. It can also be considered as an adaptive motor behaviour if a subject is incapable of making fast corrections to control the position of the CoM. The arm lifts were associated with a feed-forward CoM acceleration, directed forward in all the children with MMC and for all except one in the control group. This is in harmony with earlier studies of the GRF in standing and seated subjects (18, 52). In contrast to the results of Moore et al (52), we found no significant correlation between the onset time of CoM acceleration and the peak acceleration in either group. The children in our study made bilateral arm lifts, while in their study the subjects made unilateral arm lifts and reached as far as possible, which is a greater threat to the postural stability.

Reaching movements
The most obvious difference between the groups was reduced spatial precision in the MMC group. This observed reduction of movement precision could be explained by disturbances in the visual as well as the motor system. Harris and Wolperth (30) have discussed the impact of noise on movement performance and formulated the minimum-variance theory. They highlight that the neural signals in the sensory and the motor systems are corrupted by noise, present at every level of the sensorymotor chain and having an effect on endpoint precision. Noise in the sensory system will lead to uncertainty in the position at which the target is perceived, and noise in the motor system will affect movement co-ordination. It is likely that visual problems in subjects with MMC contributed to their poor spatial precision. Ito et al (34) have suggested that children with MMC and hydrocephalus have visuoperceptual disturbances due to dysfunction in the visual cortex and the visual pathways. The fact that the MMC group often performed better without visual FB supports this suggestion.

Impairments more directly related to the motor system could be due to Chiari's malformation and abnormalities of the brainstem and the cerebellum. It has been suggested that cerebellar and cervical cord abnormalities exert a deleterious influence on arm and hand function in children with MMC (53). The lateral brainstem pathways influence goal-directed limb movements such as reaching, though the highest level of motor control is the cortex (22). In the cerebellum motor commands are continuously updated during the course of the movement, in order to ensure movement precision (22). Both brainstem and cerebellar dysfunction could induce co-ordination problems, manifested by longer and less straight
movements in the MMC group compared to the controls. However, from our results it is not possible to distinguish the relative contribution of neuromotor impairment vs. visual processing impairment to the altered control of reaching in subjects with MMC.

According to the minimum-variance theory the central nervous system tries to minimise the consequences of noise in the systems by adapting the speed of the movement to the accuracy demands of the task. In a system with a high level of noise in the sensory or motor system, movements need to be carried out with reduced speed or the final movement precision will be low. We expected that the children with MMC would have longer movement times than the controls, but apparently with these kinds of uncomplicated hand movements they do not have a problem with movement speed. However, more complex tasks might force them to make corrective movements, resulting in prolonged movement time.

Within the MMC group subjects who had had early symptoms of brainstem dysfunction also had shorter deceleration phases compared to the others, indicating that they approached the target with higher velocity. This might contribute to inaccuracy and be perceived as clumsiness in everyday tasks. In our study, the deceleration phases were shorter during trials with NoFB, and this is in contrast with the findings of Smyth et al (75). Their subjects made reach-to-grasp trials, which are more complex than reaching only and this might explain the differences in results.

The results of this study also showed that subjects with MMC responded to the withdrawal of visual feedback in a way similar to that of the controls. Withdrawal of visual feedback during the course of the movement will normally not affect the main features of the movement, since the movement parameters are programmed in advance. However, in subjects with impaired motor programming the movement will be markedly altered if visual feedback is not available (24). This strengthens the suggestion that reduced precision was not due to impaired programming. In the literature it has earlier been shown that children with MMC and hydrocephalus have difficulty with movement planning in complex tasks, even if these planning problems were less severe than their visuospatial difficulties (17). In our study the tasks were not complex in the sense of movement planning and sequencing, and this could explain why the MMC group did not differ from the controls in movement programming. Wills (84) has also suggested that children with MMC and hydrocephalus are able to carry out simple reaction
tests but still have difficulty with tasks that demand more active screening, planning and sequencing. We have found no earlier studies investigating the movement characteristics of reaching in subjects with MMC.

Adaptation of reaching

The design of study IV was similar to that of Pine et al (58), where they investigated learning of scaling factors for reaching movements in adults. In agreement with their findings we found that movement error initially occurred after gain changes and also that movement adaptation occurred rapidly under the altered condition in subjects without disabilities. However, from our results it was obvious that reaching was not rapidly adapted to the new gain in the MMC group. Motor adaptation problems could be due to several deficits, since a novel behaviour initially requires processing in multiple motor and parietal areas (40). Structural abnormalities associated to Chiari II malformation could explain the poor motor adaptation in the MMC group. Our results indicated that the group of subjects who had had symptoms of early and severe brainstem dysfunction also had more pronounced variability in percentage error. The cerebellum is assumed to play a major role in motor adaptation, by detecting error in one movement and then adapting the motor program to the next movement (23).

The visuomotor transformation in reaching movements involves pathways from the visual cortex and the parietal areas into the premotor and primary motor areas (40). It is likely that children with MMC and hydrocephalus have problems with visual processing and visuomotor transformation, due to dysfunction of the visual cortex and the visual pathways (34). However, in this study there was no significant difference in movement error between the shunt treated and not shunt treated groups under the base condition. Furthermore, subjects who had shunt treated hydrocephalus did not show problems with motor adaptation compared to subjects without hydrocephalus. This could be due to lack of statistical power, or this might be explained by the fact that the movements were executed without visual feedback. Instead, visual information about target location and hand position were only provided before and after movement onset, which means longer time for visual processing and visuomotor transformation.

In visually guided reaching, the movement parameters are programmed in a relative co-ordinate system according to a global scaling factor (27, 40, 41, 58). Accordingly, kinematic accuracy depends on learning scaling factors
from extent errors and reference axis from directional errors, including storage and processing of movement errors in working memory (41, 58). It is possible that children with MMC have impaired working-memory function, leading to difficulties in adapting to a new visuomotor gain. Still, after the second gain change the percent error increased again also in the MMC group, which means that short-term motor learning did occur under the altered condition, even if to a lesser degree as in the control group. In trial 31, after the first and expected gain change, the MMC group had significantly larger error compared to the control group. The controls adapted their movements in advance after the verbal information about the forthcoming gain change, while subjects with MMC might not be able to rapidly process or store this information in working memory. Perhaps they need more time to produce the same motor adaptation, as do children without MMC. In trial 76, after the unexpected gain change, there was no significant difference in percentage error between the groups.

Studies of motor learning in children with disabilities are limited. However, motor adaptation to weight biasing tasks and prism adaptation tasks was recently investigated in children with MMC and children with ADHD compared to healthy siblings (10). Contrary to the authors’ expectations and to our results, they found that children with MMC did not display deficits in motor adaptation. However, there were some procedural differences that might explain the contradictory results. First, the distorting prisms caused directional errors in their study, while gain changes caused extent errors in our study. According to earlier studies, movement extent and direction have different sources of error, suggesting that they can be separately specified by the brain (27, 41). Secondly, the inclusion criteria of the studies were slightly different. Children with a history of significant neurological complications aside from hydrocephalus were not included in their study, while both subjects who had had early and severe symptoms of brainstem dysfunction and subjects without shunt treated hydrocephalus were included in our study.

General discussion and future studies
Motor problems are complex in children with MMC, and the variability within this group of children is large. This means that it can be difficult to fully understand and handle activity problems in these children. The findings of these studies contribute to the overall picture of impairments above the cere level, which may influence the motor performance. These findings also
confirm that motor impairments above the cele level are common in children with MMC as a group, which is in accordance with clinical experience and literature references. Furthermore, the associated activity problems seem to increase with age, when the demands on skilfulness in motor performance also increase. Therefore, children with MMC need regular follow-up and careful investigation of their motor performance, for the purpose of identifying and eliminating activity problems.

The clinical implication of these findings is that intervention programs are routinely adjusted for the individual child. For example, the presence of impaired posture control and stability requires guided practice of balance tasks. It also influences what kind of orthoses or technical aids are prescribed for a child. Another example is that increased independence in mobility might be achieved by increased hand strength in children who use a wheelchair. We suggest that the process of learning to move about without assistance should start early in childhood to make it easier for children with MMC to mature into well-functioning adults in the mobility domain. In addition, the parents of children with MMC require support in achieving a balance between assisting or not assisting their child. They must also be helped to set realistic expectations and to value the independence of their child. In addition, independence in daily activities often requires modifications of the environment. Sometimes, this is even a prerequisite for participation in daily activities in subjects with disabilities.

Slow motor performance is well recognised in children with MMC, involving functional limitations in daily life activities. This needs to be considered in therapy programs, together with the great issue of providing orthotic devices and technical aids for these children. A relatively natural approach to slow motor performance is to stimulate movement speed in children with MMC. However, if the slowness is a compensation for poor precision, as we found in study III, therapy programs ought to focus on movement precision instead. These children need more time than children without disabilities, to compensate for both poor movement precision and reduced capacity in motor learning. For skilfulness in daily activities it is necessary to practice a great deal, since voluntary movements improve with practice. It is also necessary to practice a variety of tasks under different visuomotor conditions. However, strategies for improving movement quality are as yet not obvious in children with MMC and impaired hand function.
There is a need for further studies to confirm these findings and to increase knowledge about motor performance in children with MMC. In a forthcoming study we will investigate the interaction between posture control and attention. The control of posture is usually regarded as independent of attention demands. However, recent research has shown that increased cognitive demands can modify posture control (12, 87). Posture control has been investigated in healthy subjects while standing on a force plate and performing different working memory (WM) tasks (12). The results showed that when a WM task was added, regardless of task type, changes in the postural sway were characterised by increased frequency and decreased amplitude of the sway. The authors concluded that the results indicated a tighter posture control strategy under the influence of a WM task. The relationship between posture control and increased cognitive demands has implications for our understanding of balance problems. By studying the effects of a WM task on the control of posture it may be possible to identify whether children with MMC are likely to be destabilised by concurrent mental stress, indicating that they are vulnerable to interference between mental activity and posture control.
Conclusions

These studies identified impairments above the cele level, which influenced mobility and sitting posture in children with MMC. In addition, the movement characteristics of reaching presented new data, which could partly explain the problems in hand activities in these children. The following conclusions were drawn from the main results:

- Most children with MMC were dependent on other individuals in daily mobility. Poor hand function and cognitive function and especially poor hand strength could make children more dependent on caregiver assistance in mobility.

- Neurological dysfunction associated with Chiari II malformation and the presence of scoliosis influenced both mobility and sitting posture.

- The low postural sway frequency in the MMC group indicated a slow motor performance. Inability to perform fast, high-frequency adjustments of the postural sway could induce posture control problems.

- The kinematic analysis of reaching movements illustrated that the slowness in hand activities could be explained by poor precision and coordination during the execution of the movements.

- Motor adaptation was impaired in subjects with MMC, even if motor learning did occur in the MMC group.

- Neurological dysfunction associated with Chiari II malformation and the presence of shunt treated hydrocephalus influenced the motor performance of reaching movements.

All these findings have implication for intervention and therapy programs for the individual child. Strategies for improving motor function of daily activities need to be further investigated in children and young adults with MMC.
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Barn med ryggmärgsbråck har vanligtvis problem med dagliga aktiviteter, men bakgrunden till deras problem är inte helt uppenbar. Förståelsen för tänkbara orsaker till aktivitetsproblem är en förutsättning för effektiv sjukgymnastik. Syftet med dessa studier var att identifiera nedsatta funktioner ovanför bråcknivån, vilka skulle kunna påverka förflyttningseffekten hos barn med ryggmärgsbråck (studie I) och att analysera hållningskontrollen i sittande (studie II) samt att analysera rörelsemönstret och kontrollen av mälriktade handrörelser (studie III & IV).

Totalt undersöktes 41 barn och ungdomar med ryggmärgsbråck och utan mental retardation. I studie I ingick 32 barn, 6-10 år. Förflyttningseffekten och assistansbehovet i dagliga aktiviteter bedömdes enligt Pediatric Evaluation of Disability Inventory och korrelationen mellan förflyttningseffekten och neurologiska avvikelser, handfunktion och kognitiv funktion beräknades. Resultatet visade att nio barn kunde förflytta sig självständigt och att det fanns en mätlig, statistiskt signifikant korrelation mellan stort assistansbehov och hög bråcknivå, nedsatt handfunktion och nedsatt kognitiv funktion. Hos de barn som förflyttade sig med rullstol var endast nedsatt handstyrkan signifikant korrelerad med ökat assistansbehov.

precision, krokigare rörelsesträcka och kortare uppbromsning av rörelsen jämfört med kontrollerna. I båda grupperna var målriktade handrörelser förprogrammerade. Gruppen med ryggmärgsbråck kunde sämre anpassa sina handrörelser till en ny visuomotorisk betingelse, jämfört med kontrollgruppen.

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