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ARTIGO ORIGINAL

Acompanhamento da locomoção de pacientes com mielomeningocele da Associação de Assistência à Criança Deficiente (AACD) em São Paulo – SP, Brasil

Ambulation follow-up in patients with myelomeningocele treated at the Associação de Assistência à Criança Deficiente (AACD) in São Paulo, Brazil

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RESUMO

Introdução: a Mielomeningocele (MMC) é um tipo de malformação congênita da coluna vertebral e medula espinhal, caracterizada por paraplegia flácida e alteração sensitiva abaixo do nível da lesão, acompanhada de comprometimento neurológico, urológico e ortopédico. Os pacientes podem ser classificados funcionalmente como torácicos (T), lombares altos (LA), lombares baixos (LB) e sacrais (S) ou assimétricos. Objetivo: traçar o perfil dos pacientes atendidos na clínica de MMC da AACD - SP considerando variáveis relacionadas ao padrão de marcha. Método: revisão dos prontuários de pacientes atendidos em avaliação inicial durante o ano de 2000, com idade inferior a um ano, e suas evoluções até última consulta na clínica no ano de 2004. Resultado: no total passaram 230 pacientes em avaliação inicial na clínica de MMC da AACD - SP no ano de 2000. Destes, 64 (27%) apresentavam menos de 1 ano de idade na primeira consulta. Destes, 11% não retornaram em consulta médica na clínica após a avaliação inicial, e dois pacientes sabidamente evoluíram para óbito. A média de idade no último retorno na clínica foi de 3,5 anos. Ao analisarmos o nível neurológico no retorno encontramos 43% nível Torácico, 20% nível Lombar alto, 28% nível Lombar baixo, 2% nível Sacral e 6% Assimétrico. Ao analisarmos o padrão de marcha observamos que 57% são não deambuladores, 7% são deambuladores não funcionais, 25% são deambuladores domiciliares e 11% são deambuladores comunitários. Entre todos os pacientes deambuladores a idade de início da marcha foi em média 3 anos. Sabe-se que pacientes com níveis neurológicos mais baixos tendem a manter a marcha por mais tempo. Como esses pacientes tendem a se tornar menos ativos e perder a marcha com o passar dos anos (devidosobretudo à obesidade e deformidades ortopédicas), é fundamental estudar a idade de aquisição da marcha. Ao analisarmos a presença de deformidades ortopédicas em coluna observamos que 57% não apresentam deformidades, 9% apresentam escoliose toracolombar, 32% apresentam cifose e 1% apresenta hiperlordose. Medula presa ocorreu em 36%. Conclusão: os níveis funcionais mais altos estão associados à aquisição mais tardia da marcha, bem como mais deformidades ortopédicas e maior necessidade de meios auxiliares.

PALAVRAS-CHAVE

criança, mielomeningocele, marcha, reabilitação, centros de reabilitação

ABSTRACT

Introduction: Myelomeningocele (MMC) is a congenital malformation of the vertebral column and spinal cord, characterized by flaccid paraplegia and sensitive alteration below the lesion level, accompanied by neurological, urological and orthopedic impairment. The patients can be classified as thoracic (T), high lumbar (HL), low lumbar (LL) and sacral (S) or asymmetric. Objective: to describe the profile of the patients treated at the MMC Clinic of the Assistance Association to the Defective Child - AACD – SP, considering variables related to the gait pattern. Methods: review of files of the patients seen at the initial assessment during the year 2000, aged younger than one year and their follow-up until their last visit to clinic in the year 2004. Results: A total of 230 patients were seen at the initial assessment at the MMC Clinic of AACD - SP in the year 2000. Of these, 64 (27%) were younger than 1 year at the first assessment. Of these, 11% did not return to the Clinic after the initial assessment and two patients died. Mean age at the last visit to the Clinic was 3.5 years. When the neurological level at the return assessment was analyzed, we found: 43% of Thoracic level, 20% of High Lumbar level, 28% of Low Lumbar level, 2% Sacral level and 6% Asymmetric. When the gait pattern was analyzed, we observed that 57% of them were not ambulators, 7% were non-functional ambulators, 25% were home ambulators and 11% were community ambulators. Among the ambulating patients, the average age at the start of gait was 3 years. It is known that patients with lower neurological levels tend to

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maintain the gait for longer periods. As these patients tend to become less active and lose gait ability with the passing years (mainly due to obesity and orthopedic deformities), it is essential to study the age at acquisition of gait ability. When we analyzed the presence of orthopedic deformities in the column, we observed that 57% of the patients did not present deformities, 9% presented thoracolumbar scoliosis, 32% presented kyphosis and 1% presented hyperlordosis. Tethered spinal cord syndrome occurred in 36% of the patients. Conclusion: The higher functional levels are associated to a later acquisition of gait, as well as more orthopedic deformities and higher need for auxiliary means.

KEYWORDS
child, meningomyelocele, gait, rehabilitation, rehabilitation centers

INTRODUCTION

Myelomeningocele (MMC) is a congenital malformation of the vertebral column and spinal cord, characterized by flaccid paraplegia and sensitive alteration below the lesion level, accompanied by neurological, urological and orthopedic impairment.[1] Other general problems can occur, such as obesity (increased food ingestion and decreased energy consumption), pressure ulcers (bed sores - areas of hyperpressure on insensitive skin), constipation and/or fecal incontinence and sexual dysfunction in adult patients. Survival during the first year of life for these patients increased in recent years from less than 10% to more than 90%, with intervention procedures such as the early closing of the defect and the ventriculoperitoneal shunt (VPS).[2] These measures, associated to the better treatment of the neurogenic bladder, have made it possible for these patients to reach adult life.

The neurological problems include, among others, the myelomeningocele sac (exposed nervous tissue covered by epidermis), obstructive hydrocephaly (in 90% of the cases[1] caused by tamponment of the foramen magnum by the cerebellum amygdala) and tethered cord. Due to malformations in the lumbar region associated to the scar caused by the surgical closing of the sac, the terminal edge might remain fixed at L5 or S1 levels (instead of being situated at L1 level) and the spinal cord is progressively stretched during the child's development phase. The most common symptoms include the onset or worsening of scoliosis, alterations in the gait pattern, alterations in the micturition pattern and spasticity.

Orthopedic deformities can occur in the trunk (kyphosis, scoliosis and hyperlordosis). Kyphosis is a severe deformity, which is often seen in the lumbar region and worsens with growth, resulting in important functional limitations. Scoliosis affects up to 50-90% patients in some studies.[3] Lumbar hyperlordosis is mainly caused by the tethered cord and this deformity can be masked by obesity and hip flexion contractures.

Hip deformities: flexion-abduction (patients at the thoracic level due to the habit-forming abandon attitude, may be associated to knee flexion), flexion-adduction (patients at the high lumbar level due to the isolated action of the flexor and adductor musculature), uni- or bilateral hip dislocation (can occur in up to 40% of the cases).[4] Knee deformities: flexion, curved (less frequent), varus, valgus. Ankle deformities: valgus (in 85% of the cases). Foot deformities: equinovarus (more common), calcaneovalgus (patients at the low lumbar level due to the isolated action of the dorsiflexors) and talipes equinus (rare).

Hoffer classified the MMC in functional levels according to the neurological impairment: thoracic (T), high lumbar (HL), low lumbar (LL) and sacral (S).[5] The prognosis of ambulation and the objectives to be attained at rehabilitation depend not only on the neurological level, but also on the presence or not of orthopedic deformities, obesity, decreased cognitive skills and the family's socioeconomic conditions.[6]

Gait, which is the most studied functional aspect, is usually classified in four types: community (610 m), home (30 m), non-functional (<3 m) and non-ambulator. The thoracic level has a bad prognosis for gait, as the patient does not present an active mobilization of the lower limbs. If the patient does not have orthopedic deformities, the objective of rehabilitation is to attain the use of well-adapted wheelchair, orthostatism with dynamic parapodium or therapeutic gait with long-leg brace + RGO (reciprocal gait orthosis) and walker.

The high lumbar level has a prognosis of regular gait, as the patient has functioning psoas, adductor and eventually quadriceps muscles. To maintain the home gait, the patient needs weight maintenance, correction of existing orthopedic deformities and the use of long orthosis with a pelvic belt and walker or crutches. At the adult age, the individual usually choose the wheelchair.

The low lumbar level has a good gait prognosis, as the patient has functioning psoas, adductor, quadriceps, medial knee flexor and eventually anterior tibial and/or gluteus medium muscles. The patient needs a suropodalic orthosis and a gait aid.

The sacral level has very good gait prognosis, as the patient has all the above mentioned functioning muscles and also plantar flexion function and/or hip extensor function. Some patients do not need an orthosis to ambulate (in these cases, it is important to verify the possibility of the individual presenting plantar perforation).

As the patients with MMC tend to become less active and stop ambulating with the passing of years, due mainly to obesity and orthopedic deformities, it is essential to study the age of gait acquisition and loss. It is known that patients with lower neurological levels tend to maintain gait for longer periods.

OBJECTIVE

The objective of the present study was to delineate the profile of the patients treated at the MMC Clinic of the Assistance Association to the Defective Child - AACD – SP, observing the initial condition at their arrival at the Clinic and their evolution up to the last medical visit, aiming at determining the gait pattern acquired by these patients.

METHOD

The medical files of all patients who were initially evaluated at the MMC Clinic of AACD-SP during the year 2000 were analyzed. Of these, we selected the patients that were less than one year of

age at the date of the first medical visit. Medical visits usually take place every 4 to 6 months at the clinic. We used the last medical visit during the year 2004. Any intercurrent event or failure in filling out the medical file was considered to be exclusion criteria.

In this retrospective study, the data were obtained from the medical files through a research protocol. Among the data collected are: personal information, neurological level, neurological, orthopedic and urological evolution, use of orthosis and gait pattern.

RESULTS

A total of 230 patients were assessed at the initial evaluation in the year 2000. Of these, 64 (27%) were less than one year old at the initial evaluation and two patients met the exclusion criteria and were removed from the study.

Of the remaining 62 patients, 7 (11%) did not return to the medical appointment at the MMC Clinic after the initial evaluation. Of the remaining 55 patients, 2 died and the cause of death is not disclosed in the patients' medical files. Thus, 53 patients remained and were enrolled in the present study.

The mean age at the last visit to the Clinic in 2004 was 3.5 years. When the neurological level was analyzed, we observed that 44% (23 patients) presented the thoracic level (T), 20% (11 patients) presented high lumbar (HL), 28% (15 patients) presented low lumbar (LB), 2% (1 patient) presented sacral level (S) and 6% (3 patients) had asymmetric neurologic involvement (different combinations of neurological levels between the right and left hemibodies, with 1 patient HL/LL and 2 patients HL/T).

When the gait pattern was analyzed, we observed that 57% (30 patients) did not ambulate (NA), 7% (4 patients) were non-functional ambulators (NFA), 25% (13 patients) were home ambulators (HA) and 11% (6 patients) were community ambulators (CA). When they were separated according to the neurological level we observed that:

Table 1
Gait pattern according to the neurological level in individuals with myelomeningocele.

	Thoracic	High Lumbar	Low Lumbar	Sacral	Asymmetric	Total
NA	21 (92%)	3 (27%)	3 (20%)	0	3 (100%)	30 (57%)
NFA	1 (4%)	2 (18%)	1 (6%)	0	0	4 (7%)
HA	1 (4%)	6 (55%)	6 (40%)	0	0	13 (25%)
CA	0	0	5 (34%)	1 (100%)	0	6 (11%)

Legend: NA non-ambulator; NFA non-functional ambulator; HA home ambulator; CA community ambulator.

Among the ambulators, the age at the onset of gait was on average 3 years. When this age was analyzed according to the neurological level, we observed that the average was 3.8 years for the Thoracic level, 3 years for the high lumbar level and 2.4 years for the low lumbar level.

We also observed that all patients with the Thoracic level had a wheelchair (around 50% of them were not adapted due to the high cost of adaptations) and the two that ambulated needed long leg brace + RGO (reciprocal gait orthosis) + walker or Canadian crutches.

Patients with high lumbar level also had a wheelchair for long distances and all those who ambulated needed the help of a long leg brace with or without thoracic extension, in addition to a walker or Canadian crutches.

Of the three patients with low lumbar level who did not acquire gait, 1 patient presented obesity and orthopedic deformity in the lower limbs and was awaiting orthopedic surgery at the institution, 1 patient was in the immediate postoperative period of orthopedic surgery and 1 patient was awaiting the manufacturing of a leg brace to start the gait training. Of the 12 ambulating patients with low lumbar level, 5 needed the aid of a leg brace and walker or Canadian crutches (being 1 CA, 2 HA and 1 NFA) and 7 needed a suropodalic orthosis with or without a lateral rod and walker or Canadian crutches (being 3 CA and 4 HA).

When we analyzed the presence of orthopedic deformities in the spinal column we observed that 30 patients (57%) did not present any deformities. Among those who presented deformities there were: 5 patients (9%) with thoraco-lumbar scoliosis, 17 patients (32%) with kyphosis and 1 patient (1%) with hyperlordosis. Table 2 shows the correlation between the spinal column deformity and the functional level according to Hoffer's classification:

Table 2
Spinal column deformities according to the functional level in individuals with myelomeningocele.

Deformity	Thoracic	High Lumbar	Low Lumbar	Sacral	Asymmetric	Total
Absent	6 (26%)	9 (82%)	14 (93%)	1 (100%)	0	30 (57%)
Kyphosis	14 (61%)	0	0	0	3 (100%)	17 (32%)
Scoliosis	2 (9%)	2 (18%)	1 (7%)	0	0	5 (9%)
Hyperlordosis	1 (4%)	0	0	0	0	1 (1%)

The patient at the sacral level did not present orthopedic deformities in the lower limbs. Among the 15 patients with low lumbar level, 4 (27%) did not present deformities and of the 11 who presented deformities there were: 9 patients (60%) with foot deformities, being 7 equinovarus and 2 calcaneovalgus, 5 patients (33%) with ankle deformity – increased external tibial torsion, 2 patients (13%) with knee flexion and 7 patients (47%) with hip flexion, of whom 3 patients presented unilateral hip subluxation.

Among the 11 patients with high lumbar level, 8 (73%) presented foot deformities in equinovarus, 1 (9%) presented knee flexion and 7 (64%) hip flexion (5 with hip subluxation). Among the 23 patients with the thoracic level, 10 (44%) presented equinovarus feet, 9 (39%) knee flexion and 15 (65%) hip flexion-adduction, of whom 5 presented hip subluxation.

Only 8 patients (15%) did not have a ventriculoperitoneal shunt (VPS). Among the patients with VPS, it was observed that 29 (65%) did not have a valve replacement up to the end of this study and among those who had, 10 patients (22%) had one replacement, 2 (4%) had 2, 1 (2%) had 3, 1 (2%) had 4 and 1 (2%) had

six replacements.

When the picture of tethered spinal cord syndrome was analyzed, it was observed that this condition occurred in 19 patients (36% of the total). Of these, 10 patients had the characteristic clinical picture associated to the confirmation by magnetic resonance imaging (MRI) results and urodynamic testing and 6 of these patients had been submitted to the surgical release of the tethered cord; it is important to remember that the functional level can correspond to more cranial medullary neurological levels after this surgery.

Only six patients (11%) were not followed by an urologist at the institution. Of the 47 remaining patients, 34 of them (72%) were initially advised to undergo Crede's maneuver every 2 hours with the objective of attaining an adequate vesical emptying; 5 patients (10%) presented reflex micturition, 7 patients (15%) had been submitted to vesicostomy due to an important vesicoureteral reflux and 1 patient (2%0 had been submitted to vesical dilation.

DISCUSSION

A study at the Vall d´Hebron Hospital in Spain,[4] with 322 patients, showed that the patients at the thoracic level initiate gait at around 6 years of age, whereas those at the lumbar level do it at around 3.5 years and those at the sacral level at 2 years. Of these patients, the thoracic ones lost ambulation at around 10 years of age, the lumbar ones at around 15 and the sacral ones continued to ambulate. The reasons for the loss of ambulation included orthopedic deformities, obesity and high energy consumption necessary to ambulate with the long leg brace. However, there is a difference in the criteria of functional classification. When the gait pattern was analyzed, it was observed that 22% were NA, 3% were NFA, 10% were HA and 65% were CA. When the neurological level was considered, 96% of the sacral patients showed to be community ambulators and 94% of the thoracic patients were non-ambulators, which is similar to the data found in the present study.

As for the orthopedic deformities in the lower limbs and column, it was observed that in 27% of the cases there is hip flexion, being more frequent for the T and HL levels, due to the muscular imbalance between the flexor and extensor muscles of the hips. In 39% of the cases there is uni- or bilateral hip subluxation, with conservative treatment due to the fact that it is nonpainful hip. The column deformities are present in 45% of the patients, with scoliosis being the most frequent (89%), with surgical treatment in 18% of the cases, followed by kyphosis (10%).

A study carried out at the Shriner's Hospital in the United States showed for the thoracic level that the percentage of patients that ambulated until the adolescence varied from 0 to 33%. For the High Lumbar level, it varied from 10 to 54%, for the Low Lumbar level, it varied from 31 to 95% and for the sacral level, it varied from 53 to 100%.[6]

As our institution is a Reference Center in the treatment of MMC and due to the fact that we treat patients with higher neurological levels (43% thoracic in relation to 2% sacral), we have a total of 57% of patients who do not ambulate and 11% of community ambulators, with the age at the onset of gait being around 3 years. As for the orthopedic deformities, it is known that they are related to the functional level of myelomeningocele and it was observed in our study that 50% of the total number of patients presented foot deformities (more frequent at the LL level), 10% presented ankle deformities, 22% presented knee deformities and 55% presented hip deformities (more frequent at the HL and T levels).

The orthopedic deformities have an impact on these patients' functionality and can impair orthosis use and gait onset. For the patients with a gait prognosis, the surgical correction of these deformities is indicated; for those without a gait prognosis, the surgical approach has an esthetic aim and it is not carried out routinely at our Service.

CONCLUSION

The present study showed that the functional level is related to the age at gait onset and pattern and that the patients with the higher functional levels acquire the gait later, with the use of more auxiliary devices and that they present orthopedic deformities more frequently. On the other hand, the patients with lower functional levels had fewer deformities, needed fewer auxiliary devices and acquired gait earlier.

These facts were in agreement with the data found in the literature regarding the age at the gait onset and gait pattern in patients with MMC, which warrants further studies in order to determine when the gait loss occurs in these patients.

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