

SPINAL MUSCULAR ATROPHY TYPE II (INTERMEDIARY) AND III (KUGELBERG-WELANDER)

EVOLUTION OF 50 PATIENTS WITH PHYSIOTHERAPY AND HYDROTHERAPY IN A SWIMMING POOL

MÁRCIA C. B. CUNHA*, ACARY S.B. OLIVEIRA**,
RITA HELENA D.D. LABRONICI***, ALBERTO ALAIN GABBAI****

ABSTRACT - We added hydrotherapy to 50 patients with spinal muscular atrophy (SMA) who were being treated with individual conventional physiotherapy. Hydrotherapy was performed at an approximate temperature of 30 degrees Celsius, twice a week, for thirty minutes in children and for forty-five minutes in adults during a 2-year period. The outcome derived from this combined modality of treatment was rated according to physiotherapeutic evaluations, the MMT (Manual Muscular Test), and the Barthel Ladder. Patients were re-evaluated at 2-month intervals. After two years of ongoing treatment, we were able to observe that the deformities in hip, knee and foot were progressive in all SMA Type II patients, and in some Type III. Muscle strength stabilized in most SMA Type III patients, and improved in some. MMT was not done in SMA Type II. In all patients we were able to detect an improvement in the Barthel Ladder scale. This study suggests that a measurable improvement in the quality of daily living may be obtained in patients with SMA Types II and III subjected to conventional physiotherapy when associated with hydrotherapy.

KEY WORDS: physiotherapy, hydrotherapy, spinal muscular atrophy.

Atrofia muscular espinhal tipo II (intermediária) e III (Kugelberg-Welander): evolução de 50 pacientes com fisioterapia e hidroterapia em piscina

RESUMO - A hidroterapia foi realizada em 50 pacientes com atrofia muscular espinhal, os quais foram também tratados com fisioterapia individual convencional. O tratamento hidroterápico foi realizado em piscina aquecida numa temperatura de aproximadamente 30° Celsius, duas vezes por semana, durante 30 minutos em crianças e 45 minutos em adultos num período de dois anos. Os benefícios deste tipo de tratamento foram avaliados de acordo com a evolução clínica, o MMT (Teste de Força Muscular) e a Escala de Barthel. Os pacientes foram reavaliados a cada dois meses. Após dois anos de tratamento nós observamos que as deformidades nos quadris, joelhos e pés foram progressivas em todos os pacientes do Tipo II e em alguns do Tipo III. Houve estabilização da força muscular na maioria dos pacientes com SMA Tipo III, e melhora da força em alguns; nos pacientes com SMA Tipo II este teste não pode ser realizado. Em todos os pacientes pudemos verificar melhora em relação à Escala de Barthel. Este estudo sugere que houve melhora destes pacientes com SMA Tipos II e III em relação às atividades de vida diária quando a fisioterapia convencional foi associada a hidroterapia.

PALAVRAS-CHAVES: fisioterapia, hidroterapia, atrofia muscular espinhal.

Spinal muscular atrophy (SMA) manifests as diffuse muscle weakness secondary to degeneration of the anterior horn cells and bulbar nuclei without pyramidal tract involvement^{5,6,7}.

* Fisioterapeuta, Mestre em Neurociências Universidade Federal de São Paulo - Escola Paulista de Medicina (UNIFESP-EPM); **Doutor em Neurologia, UNIFESP-EPM; ***Mestranda em Neurociências UNIFESP-EPM; ****Livre Docente em Neurologia UNIFESP-EPM. Aceite: 6-março-1996.

There are at least 3 types of SMA¹⁹. Those which affect children include SMA I, a serious condition (Werdnig-Hoffmann disease), and SMA II (intermediary form). SMA III (Kugelberg-Welander) is a more benign form which becomes evident in late childhood or adolescence^{1,15,22}. In spite of this heterogeneity all of the different forms are caused by the same genetic error on chromosome 5 (5q)^{3,18}. It is certain that most, if not all, of the patients with SMA have a progressive motor deterioration, in spite of conventional physiotherapeutic treatment⁴.

The main purpose of this study was to evaluate the potential benefits of hydrotherapy as a supplementary rehabilitation method for SMA Types II and III patients.

METHOD

We treated 50 patients with SMA Types II (intermediary) and III (Kugelberg-Welander). The diagnosis was based on clinical presentation, EMG study and muscle biopsy, according to standard criteria. Thirty patients were Type II, 17 males (56,6%) and 13 females (43,3%); and 20 were Type III, 11 males (55%) and 9 females (45%). For Type II patients, ages varied from 1 year and 10 months to 10 years and 1 month (mean = 2.9 years; median = 2,3 years); for Type III patients, ages varied from 6 to 40 years (mean = 18 years; median = 15 years).

Patients were examined every 2 months always by the same professional (MCBC) who also performed some of the hydrotherapy sessions. Parameters included: a) deformities in joints^{1,5,6}; b) development of scoliosis^{21,24,25}; c) evolution of muscular strength (Manual Muscular Test, MMT)^{7,24}; d) Barthel Ladder¹⁷; and e) motorial activities^{20,22}.

Patients were treated for 2 years with individual physiotherapy once a week, and hydrotherapy and therapeutic swimming (Halliwick method)^{10,12,13,16,23,26} in a heated swimming pool at an approximate temperature of 30C twice a week. Hydrotherapy lasted 30 minutes for children and 45 minutes for adults. Individual physiotherapy consisted of cinesiotherapy with respiratory exercises⁸, and stretching².

RESULTS

Table 1 shows that the degrees of deformities in the hips, knees and feet increased in all 50 patients during the 2-year interval of our study.

Table 2 shows that the degrees of scoliosis in our Type II patients was more pronounced than in Type III, and as the disease progressed, in spite of the treatment there was an increase in the overall number of patients who developed scoliosis and kyphosis.

Only Type III patients were analysed with the MMT since that test is not reliably performed in small children. Table 3 shows that muscle strength either stabilized or improved in this group of patients.

Table 4a shows that 93% of Type II and 100% of Type III patients improved their daily activities (Barthel Ladder) after 2 years of treatment. Table 4b shows in detail the specific areas of improvement.

Table 5a shows that Type II patients improved their motorial activities except for walking. Four patients lost their walking capacity during the treatment. Type III patients improved all motorial activities after the treatment including walking (Table 5b).

DISCUSSION

1. Physiotherapeutic aspects

There are limited data on physiotherapeutic and hydrotherapeutic treatment applied to SMA patients. In 1986, Bussi and Mangosto mentioned the importance of individual physiotherapy, respiratory and "thermo-hydrotherapy", for SMA patients, applied for 20 minutes in a swimming pool heated at 30C. The authors did not describe objective results, but mentioned the beneficial psychological effects that the water has brought to their patients⁴.

We applied individual physiotherapy and hydrotherapy in 50 patients for 2 years. Since gravity has no effect in water, our patients were able not only to float but also to accomplish a larger range of movements. Subjectively we think that the psychological effects were also remarkable. Patients felt the freedom to move around, the opportunity to unleash themselves emotionally, and the feeling of success and accomplishment. These factors led to an improvement in their physical condition and well-being.

Table 1. Evolution of deformities in hip, knees and feet in 50 patients with SMA Types II and III, before and after treatment.

Degrees of deformity	Hip		Knees		Feet	
	B	A	B	A	B	A
Mild	40	22	40	21	48	43
Moderate	10	20	10	22	2	6
Severe	0	8	0	7	0	1
Total	50	50	50	50	50	50

Mild from 5° to 20°; Moderate from 20° to 40°; Severe > 40°. B, before treatment; A, after treatment.

Table 2. Evolution of scoliosis and kyphosis in 50 SMA Type II and III patients, before and after treatment.

Deformities	Type II		Type III	
	B	A	B	A
Scoliosis	12	18	2	8
Kyphosis	9	12	12	14

B, before treatment; A, after treatment

Table 3. Evolution of muscle strength (MMT) in 20 SMA Type III patients after treatment.

	Upper Limbs		Lower Limbs	
	Proximal	Distal	Proximal	Distal
Same	12	16	7	14
Better	8	4	13	6
Worse	0	0	0	0

MMT, Manual Muscular Test.

Table 4a. Evolution of patients according to the Barthel Ladder after treatment.

Evolution	SMA Type II	SMA Type III	Total
Improvement	28	20	48
Same	2	0	2
Worsening	0	0	0
Total	50	50	50

Table 4b. Improvement in specific daily activities according to the Barthel Ladder after treatment.

	SMA Type II		SMA Type III	
	Male	Female	Male	Female
Patients	17	13	11	09
Food	12	12	4	2
Bath	2	1	4	0
Toilet	11	9	0	5
Dressing	11	9	6	4
Use of toilet bowl	10	11	2	3
Transfers bed/chair	6	7	6	5
Walking	2	0	2	1
Stairs	0	0	2	1

Table 5a. Motorial activities accomplished by SMA Type II patients, before and after physiotherapeutic and hydrotherapeutic treatment.

Motorial activities	Rolling	Sitting	Standing	Walking
Before	30	25	13	7
After	30	30	17	3

Table 5b. Motorial activities accomplished by SMA Type III patients, before and after physiotherapeutic and hydrotherapeutic treatment.

Motorial activities	Rolling	Sitting	Standing	Walking
Before	19	19	16	16
After	20	20	19	18

2. Deformities

In our present study we showed, in spite of physiotherapeutic and hydrotherapeutic treatment, that almost all patients experienced deformities worsening mainly in the hips, knees and feet. Patients with SMA Type II were the most affected developing more deformities than Type III. We did not notice any deformity in the upper limbs.

Spinal deformities were also frequent in our patients. Scoliosis and kyphoscoliosis after age 3 and collapsing kyphosis before that age are the usual patterns, with most deformities appearing by age 6²¹. Once the spinal deformity appears, progression without prevention is almost certain⁶. Several authors, including Schwentker and Gibson²⁴, have emphasized the potential for loss of walking ability and/or head control following operative intervention for these spinal deformities. With our physiotherapeutic and hydrotherapeutic treatment we think we were able to retard the progression of those deformities. None of our patients had the need to undergo surgery corrective of scoliosis.

3. Muscular strength

We noticed that with the treatment, SMA Type II patients were able to better stabilize their posture, and to better perform rolling and sitting, showing a possible muscular strength stabilization. Type III patients that were periodically evaluated through the MMT, showed stabilization and even a certain improvement. This improvement was observed mainly in the proximal portions of their lower limbs.

4. Barthel Ladder and Motorial Activities

That scale and those activities were used pre- and post-treatment to evaluate its benefits against the patients' routine activities.

It is probable that the observed improvement in the performance of their activities be a consequence of the treatment. Despite their increasing deformities patients were encouraged, since they were able to sustain their muscular strength and hence better perform their routine daily activities.

CONCLUSIONS

After a 2-year physiotherapeutic and hydrotherapeutic treatment on SMA Types II and III patients, we could conclude that:

1. Deformities in hips, knees and feet were progressive, even after the treatment. We noticed that, in SMA Type II patients, the progression of contractures were faster than in SMA Type III patients.

2. Fifty-two percent of our patients were affected by scoliosis. Twenty-four percent developed the disease during the treatment but none of them has undergone surgery. Kyphosis also affected 52% of the patients but only 10% developed the disease during the treatment period.

3. Muscular strength stabilization occurred in Type II patients, and even a certain improvement was observed in Type III. Such improvement was observed in the proximal portions of the lower limbs.

4. According to the Barthel Ladder, and the Motorial Activities, 93.3% of SMA Type II patients and 100% of Type III patients showed improvement. The items that achieved better results were food, dressing, bath, toilet, use of toilet bowl, and bed to chair transfers.

5. The hydrotherapy we introduced as a form of supplementary treatment, has brought physical and psychological benefits to SMA Types II and III patients.

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