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Contractures of the Upper Extremities in Spinal Muscular Atrophy Type II. Descriptive Clinical Study with Retrospective Data Collection

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SUMMARY

Background. The most significant orthopaedic problem for patients with the intermediate form of spinal muscular atrophy, SMA type II, is the appearance of contractures in addition to progressive scoliosis and pelvic obliquity with increasing loss of sitting stability. This study deals with restrictions of the passive range of motion and the development of contractures in the joints of the upper extremities in these patients.

Material and methods. We followed up 143 patients, 74 female and 69 male, with SMA type II for an average of 5.3 years (±4.0, 0.2 – 18.7). Their average age at the first examination was 8.4 years (±6.6, 0.1 – 34.1) and at the last examination 12.3 years (±6.5, 0.7 – 37.1). The passive range of joint motion was determined using a goniometer. According to Johnson et al. (1992), we calculated the relative contracture index (CI).

Results. The loss of range of motion and the contractures of the joints of the upper extremities (shoulder, elbow and wrist) increased progressively with age. The most marked restriction of motion was in the elbow joint with severe flexion contractures in some cases.

Conclusion. The findings of this study give us more information about the development of contractures of the upper extremities and aim to help to improve the quality of orthopaedic care of patients with SMA type II.

Key words: spinal muscular atrophy, contracture, range of motion, upper extremities, contracture index.

BACKGROUND

The onset of the intermediate form of spinal muscular atrophy (SMA type II) usually occurs before 18 months of age. The milestones of normal movement or development are never completely achieved. Predominantly, there is symmetrical muscular weakness, mostly in the proximal muscles, muscular hypotonia and hyporeflexia, frequently combined with fine tremor of the fingers. The children are able to sit on their own but will never achieve the ability to stand or walk independently. Occasionally, they learn to stand with the support of assistive devices. In many cases the clinical course is marked by stable periods of varying lengths when the disease is arrested [1]. In their study of 240 patients with SMA type II, Zerres et al. [1] indicate a survival rate of 98.5% for 5 years, 82.8% for 15 years and 68.5% for 25 years.

The fundamental orthopaedic problems for patients with SMA type II are muscular imbalance with contractures of the lower and upper extremities, sitting instability as a result of progressive scoliosis, and pelvic obliquity [2]. Fractures also occur more frequently in this population due to osteoporosis caused by inactivity [3-4]. It is a disorder with diffuse general weakness, most prominent in the lower extremities and proximal muscles, but the distal muscles and upper extremities are also affected. Functional restrictions are generally first to appear in the lower extremities, on fewer occasions they appear simultaneously in both the upper and lower extremities and in only a few patients do they appear only in the upper extremities. Restrictions of range of motion occur more frequently and usually with greater severity in the lower extremities than in the upper extremities. Elbow and/or shoulder and wrist contractures as well as hypermobile joints (often wrists) are observed [2,5-10].

Daily activities for the upper extremities are made easier by the development of compensatory and adaptive techniques, which often occur spontaneously or are facilitated by simple assistive devices, so that surgery (for example, tendon transfer) is indicated very rarely.

The purpose of this study is to determine the development and course of contractures of the upper extremities in patients with SMA type II, following a survey of 143 patients.

MATERIAL AND METHODS

This study is a retrospectively collected data set of 143 patients with SMA type II (74 females and 69 males). These patients were examined during the regular nationwide consultations for patients with neuromuscular disorders. All patients with confirmed diagnosis of SMA with type II of SMA (patients are unable to walk but they are or were able to sit unsupported) were identified for our study. This represents the entire population of our patients. The diagnosis of SMA was confirmed by human genetics in 117 patients (82%) and by electrophysiology and muscle biopsy in 26 patients (18%).

This study includes more than one evaluation of some of the patients. Thirty-seven patients were examined once and 106 were examined a number of times, on average 6 times (±5, 2-25 times), at intervals of at least 6 months. Their average age at the first examination was 8.4 years (±6.6, 0.1-34.1 years) and at the last examination 12.3 years (±6.5, 0.7-37.1 years). The mean time of follow-up of those patients who were examined a number of times was 5.3 years (±4.0, 0.2-18.7 years).

The data are presented in a cross sectional manner. The patients are divided into age groups: 0-2, 3-5, 6-10, 11-15, 16-20, 21-30 and over 30 years of age. Listed in the age range is only one evaluation from each patient that manifested an abnormality during that period. If a patient within one of the age groups had numerous examinations, only the examination that showed the greatest loss of range of motion was taken into consideration. It was almost always the last examination during this period.

Two experienced examiners used a goniometer to assess the passive range of motion. Results were documented with the neutral-null-method and the data was evaluated with the Microsoft Access data processor programme and Excel spreadsheets.

The neutral-null-method is based on a defined anatomically normal position, the neutral or functional position, called the neutral-null position (standing upright, with hanging arms, thumbs forward, feet held closed and parallel, eyes directed forward). [11] The range of motion for every joint is described by three numbers: e.g. the range of flexion, neutral-null position (0° if achieved) and the range of extension (e.g. FL/EXT of the shoulder with fixed scapula 80°/0°/20°). If the anatomically normal position (the neutral-null position) cannot be achieved, then 0° is either in the first or in the third position, depending upon the particular movement restriction present (e.g. maximal flexion of 70° and flexion contracture of 30° of the shoulder with fixed scapula: FL/EXT 70°/30°/0°).

Shoulders were examined for abduction/adduction, flexion/extension and external/internal rotation in 90° abduction. The scapula was fixed with the hand of the examiner and the measure was taken at the point when the scapula first moved. Normal phy-

siological values were defined as [11]:

- Flexion (FL) / extension (EXT) with fixed scapula 80°/0°/20°
- Abduction (ABD) / adduction (ADD) with fixed scapula 90°/0°/20°
- External rotation (ER) and internal rotation (IR) in 90° abduction with fixed scapula 80°/0/60°.

Extension/flexion and pronation/supination were tested in the elbow. The normal physiological values are [11]:

- Extension (EXT) / flexion (FL) 10°/0°/150°
- Pronation (PRO) / supination (SUP) 90°/0°/90°.

The wrist was tested for dorsal extension and palmar flexion as well as ulnar and radial abduction [11]:

- Dorsal extension (DE) / palmar flexion (PF) 60°/0°/60°
- Ulnar abduction (UABD) / radial abduction (RABD) 40°/0°/30°.

A loss of 10° or more from the norm was considered to be a loss of range of motion.

Particularly manifest flexion contractures of the elbow and ulnar abduction contractures of the wrist were evaluated separately. We took 10° of elbow extension as full extension. A loss of range of motion of 5° or more than the neutral-null position was considered to be a contracture.

We calculated a relative contracture index (CI) according to Johnson et al. [5]. We defined the relative contracture index as the proportion of patients with a loss of range of motion (or a flexion contracture of the elbow and ulnar abduction contracture of the wrist) expressed as a percentage, multiplied by the mean maximum loss of range of motion in degrees and divided by 1,000 [5]:

$$\frac{\text{(Percentage of subjects with loss of ROM) x (Mean maximal loss of ROM in degrees)}}{1{,}000}$$

This formula results in a CI of 0 to 15, according to the number of examinations and the extent of the loss of range of motion. For example: 100% of patients with contractures and a mean maximum loss of range of motion 100°, divided by 1,000 results in a CI of 10.0.

The higher the percentage of patients with loss of ROM and the cross-sectional assessment of the mean maximal loss of ROM, the greater is the potential for contracture development. [5] According to Johnson et al. [5], the risk of developing a range of motion limitation or a contracture is high when the CI is more than 1.0, moderate when it is 0.5 to 0.9 and mild when it is lower than 0.5.

The CI of the right and left joints of the upper extremities of each patient was calculated separately and the results were displayed in diagrams for each age group. Since on average the results for right and left were comparable, they have been combined in the tables showing the loss of range of motion. The exceptions were particularly a few patients over 20 years old with little examination data. A significant correlation between age and asymmetry is not demonstrable due to a very small group of patients over 20 years of age.

This study complies with the ethical standards for human research and has been approved by the Ethics Committee of Friedrich-Alexander-University Erlangen-Nuremberg.

RESULTS

Shoulder

A limitation of ABD/ADD of the shoulder of an average of 6.9° (±7.1°, 5-60°) was found in 161 of 420 examinations (Table 1). The development of the CI across the age groups showed an almost constant progression with values between 0.2 and 0.5 (Diagram 1). The abduction and adduction decrease from the age of 20. Abduction contractures were found in 2 patients: one in the 6-10 age group (15) and one in the 11-15 age group (10°). No adduction contractures were observed.

Loss of range of motion was determined in 4.8% of the examinations for FL/EXT. The losses were between 5° and 20°, on average 12.8° (±5°) (Table 1). The contracture index shows very small values which do not exceed 0.13. The high values in the group aged 30 and above correlate with the low numbers of patients and their loss of range of motion (Table 1 and Diagram 1). Flexion remained largely unchanged throughout. Extension decreased further after the age bracket of 11-15 years. In 420 examinations, only one patient, in the 11-15-year-old group, presented a flexion contracture of 10°.

The range of motion in ER/IR tests showed losses between 10° and 80°, on average 41.9° (±18.9°), in 3.1% of 420 examinations (Table 1). The CI showed an increasing tendency from the age group of 6-10 years (Diagram 1). There was only slight limitation of shoulder mobility for external and internal rotation in a few patients. One patient in the 11-15 age group had an internal rotation contracture of 10°.

Elbow

Measurements of the loss of range of motion in the elbow joint showed progressive development of

Tab. 1. Loss of range of motion (ROM) of shoulder joint in SMA Type II. Combined data for right and left limbs

Patients' age (years)	Number of examinations	Loss of ROM present (%)	No loss of ROM (%)	Loss of ROM [°]		
				Median	Mean (s_D)	Min - Max
ABD/ADD						
0 - 2	12	1 (8.3%)	11 (91.7%)		5°	
3 - 5	71	38 (53.5%)	33 (46.5%)	5°	5.1° (0.8°)	5° - 10°
6 - 10	203	74 (36.4%)	129 (63.5%)	5°	5.9° (5.8°)	5° - 60°
11 - 15	105	38 (36.2%)	67 (63.8%)	5°	8.5° (8.1°)	5° - 45
16 - 20	16	6 (37.5%)	10 (62.5%)	5°	13.6° (15.7°)	5° - 45°
21 - 30	8	2 (25%)	6 (75%)		5° and 20°	
> 30	5	2 (40%)	3 (60%)		10° and 50°	
All examinations	420	161 (38.3%)	259 (61.7%)	5°	6.9° (7.1°)	5° - 60°
FL/EXT						
0 - 2	12	0 (0%)	12 (100%)	-	-	-
3 - 5	71	0 (0%)	71 (100%)	-	-	-
6 - 10	203	8 (3.9%)	195 (96.1%)	12°	13.1° (3.6°)	10° - 20°
11 - 15	105	9 (8.6%)	96 (91.4%)	10°	10.4° (4.6°)	5° - 20°
16 - 20	16	1 (6.2%)	15 (93.7%)		20°	
21 - 30	8	0 (0%)	8 (100%)	-	-	-
> 30	5	2 (40%)	3 (60%)		5° and 20°	
All examinations	420	20 (4.8%)	400 (95.2%)	10°	12.8° (5°)	5° - 20°
ER/IR						
0 - 2	12	0 (0%)	12 (100%)	-	-	-
3 - 5	71	0 (0%)	71 (100%)	-	-	-
6 - 10	203	2 (1%)	201 (99%)		40° and 60°	
11 - 15	105	8 (7.6%)	97 (92.4%)	40°	40.8° (21.1°)	10° - 80°
16 - 20	16	2 (12.5%)	14 (87.5%)		10° and 40°	
21 - 30	8	1 (12.5%)	7 (87.5%)		50°	
> 30	5	0 (0%)	5 (100%)	-	-	-
All examinations	420	13 (3.1%)	407 (96.9%)	40°	41.9° (18.9°)	10° - 80°

Tab. 2. Loss of range of motion (ROM) of elbow joint in SMA Type II. Combined data for right and left limbs

Patients' age (years)	Number of examinations	Loss of ROM present (%)	No loss of ROM (%)	Loss of ROM [°]		
				Median	Mean (s_D)	Min - Max
EXT/FL (Loss of normal physiological ROM – with physiological extension – 10°/0°/150°)						
0 - 2	12	2 (16.7%)	10 (83.3%)		30° and 50°	
3 - 5	61	22 (36.1%)	39 (63.9%)	30°	28.9° (6.9°)	15° - 45°
6 - 10	197	95 (48.2%)	102 (51.8%)	30°	38.1° (22.5°)	10° - 130°
11 - 15	105	81 (77.1%)	24 (22.9%)	40°	49.8° (30.8°)	10° - 155°
16 - 20	16	13 (81.2%)	3 (18.7%)	40°	50.2° (36.3°)	10° - 120°
21 - 30	8	7 (87.5%)	1 (12.5%)	65°	77.1° (25.2°)	40° - 120°
> 30	5	5 (100%)	0 (0%)	85°	75° (34.1°)	10° - 110°
All examinations	404	225 (55.7%)	179 (44.3%)	30°	44.1° (27.9°)	10° - 155°
PRO/SUP						
0 - 2	17	1 (5.9%)	16 (94.1%)		20°	
3 - 5	83	18 (21.7%)	65 (78.3%)	60°	57.8° (31.1°)	10° - 100°
6 - 10	221	61 (27.6%)	160 (72.4%)	80°	72° (31.6°)	10° - 170°
11 - 15	130	59 (45.4%)	71 (54.6%)	90°	74.1° (30.2°)	10° - 160°
16 - 20	22	11 (50%)	11 (50%)	90°	81.1° (41.8°)	20° - 160°
21 - 30	10	5 (50%)	5 (50%)	67.5°	59.4° (39.5°)	10° - 100°
> 30	6	2 (33.3%)	4 (66.7%)	10°	21.7° (20.2°)	10° - 45°
All examinations	489	157 (32.1%)	332 (67.9%)	67.5°	55.1° (14.1°)	10° - 170°

flexion contractures in some patients up to complete loss of mobility. Reduction of mobility appeared already in nearly 17% of patients aged 0 to 2 years and in 87.5% of those aged from 21 to 30. The average loss of extension of the elbow was 44.1° (±27.9°, 10°-155°) as compared to the normal value (Table 2). The contracture index increased with age from 0.6 in 0- to 2-year-olds to 6.8 in 21- to 30-year-olds (Diagram 2).

The mean values of flexion and flexion contracture show that flexion remained almost constant for years but flexion contractures continued to increase noticeably with age.

Flexion contractures of the elbow also appeared in those aged 0 to 2 years and increased steadily (Table 3 and Diagram 3).

Loss of range of motion for PRO/SUP of an average of 55.1° (±14.1°, 10-170°) was found in 157 of

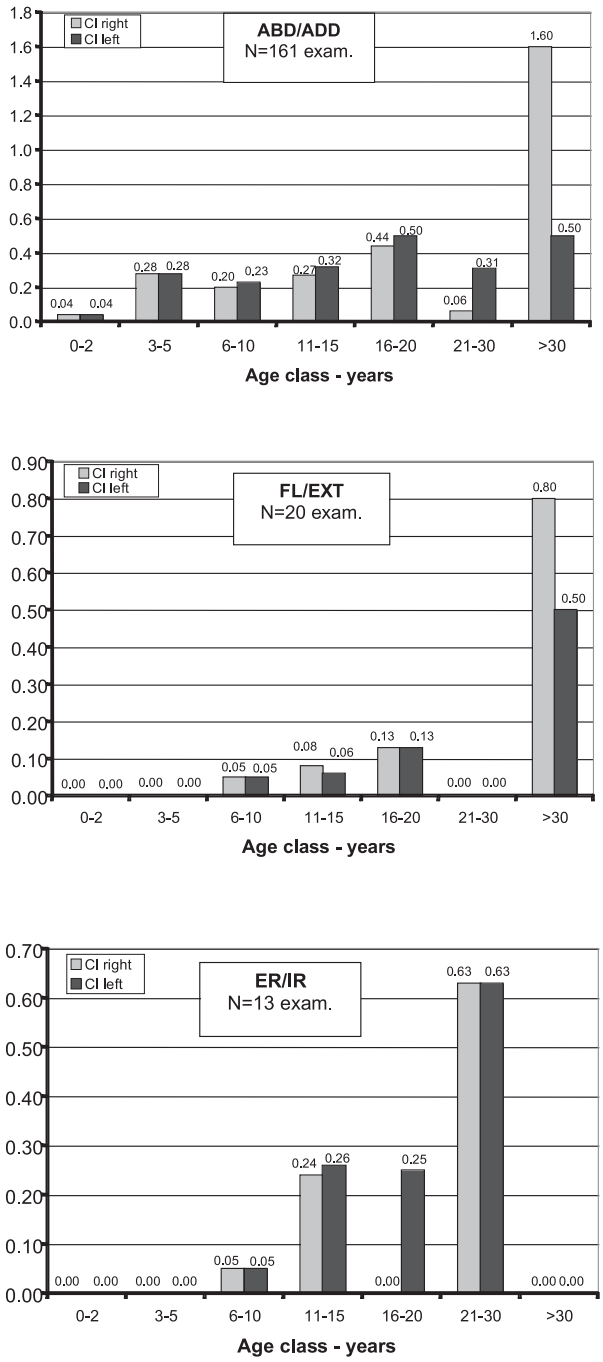

Fig. 1. Loss of range of motion (ROM) – relative contracture index (CI) – shoulder joint ABD/ADD, FL/EXT and ER/IR in SMA Type II

489 examinations (Table 2). The development of the CI across the age groups showed a progression between 1.2 for 3- to 5-year-olds to 3.5 (left) for 16- to 20-year-olds (Diagram 2). Loss of supination was observed in 157 examinations (32.1%) and loss of pronation only in 15 examinations (3.1%) (Table 4). The mean values of pronation remained mostly stable and the range of supination decreased with age.

Wrist

DE/PF tests of the wrist showed a loss of motion of an average of 41.5° (±18.7°, 10°-90°) in 7.6% of the patients. Those patients affected seemed to lose increasingly more range of motion as they grew older. However, 89% of those aged 21-30 still had unimpeded DE/PF of the wrist (Table 5 and Diagram 4). Palmar flexion remained largely unchanged over time and dorsal extension decreased slightly.

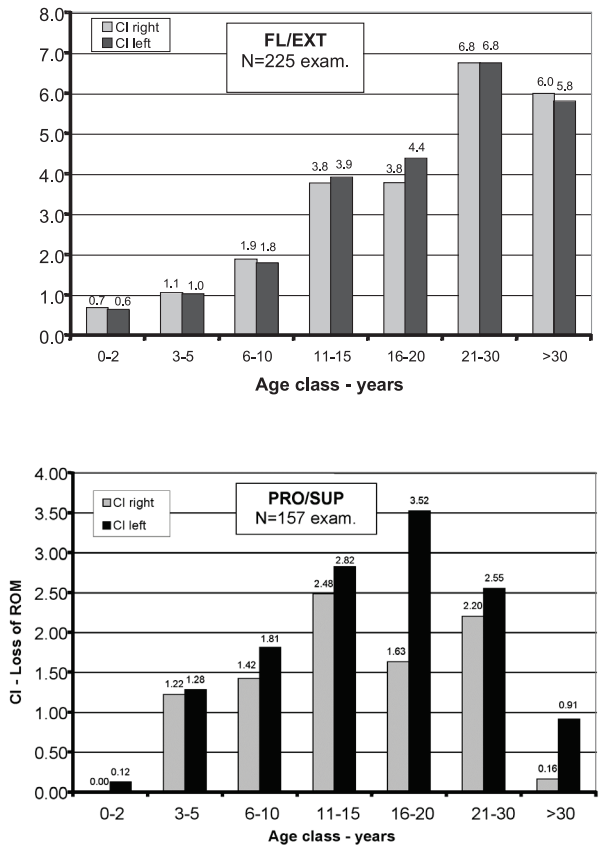

Fig. 2. Loss of range of motion (ROM) – relative contracture index (CI) – elbow joint FL/EXT and PRO/SUP in SMA Type II

Tab. 3. Manifest flexion contracture of elbow joint in SMA Type II. Combined data for right and left limbs

Patients' age (years)	Number of examinations	Contracture present (%)	No contracture (%)	Manifest flexion contracture [°]		
				Median	Mean (s_D)	Min - Max
0 – 2	12	2 (16.7%)	10 (83.3%)		5° and 10°	
3 – 5	61	7 (11.5%)	54 (88.5%)	10°	10° (5.8°)	5° - 20°
6 - 10	197	48 (24.4%)	149 (75.6%)	15°	28.1° (26.4°)	5° - 120°
11 - 15	105	70 (66.7%)	35 (33.3%)	25°	35.8° (31.5°)	5° - 145°
16 - 20	16	10 (62.5%)	6 (37.5%)	30°	42.7° (32°)	5° - 95°
21 - 30	8	7 (87.5%)	1 (12.5%)	50°	54.3° (22.8°)	30° - 90°
> 30	5	5 (100%)	0 (0%)	70°	65.6° (27.9)°	30° - 100°
All examinations	404	149 (36.9%)	255 (63.1%)	20°	34.3° (30.1°)	5° - 145°

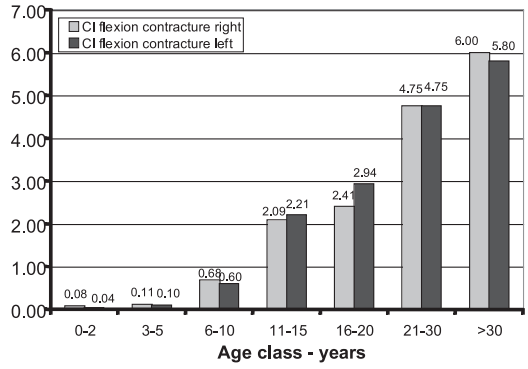

Fig. 3. Flexion contracture of elbow joint – relative contracture index (CI) – in SMA Type II. N=149 exam

Tab. 4. Loss of range of motion (ROM) of PRO and SUP of elbow joint in SMA Type II. Combined data for right and left limbs

Patients' age (years)	Number of examinations	Loss of ROM on SUP present (%)	Loss of ROM on PRO present (%)
0 - 2	17	1 (5.9%)	0
3 - 5	83	18 (21.7%)	0
6 - 10	221	61 (27.6%)	3 (1.3%)
11 - 15	130	59 (45.4%)	10 (7.7%)
16 - 20	22	11 (50%)	1 (4.5%)
21 - 30	10	5 (50%)	1 (10%)
> 30	6	2 (33.3%)	0
All examinations	489	157 (32.1%)	15 (3.1%)

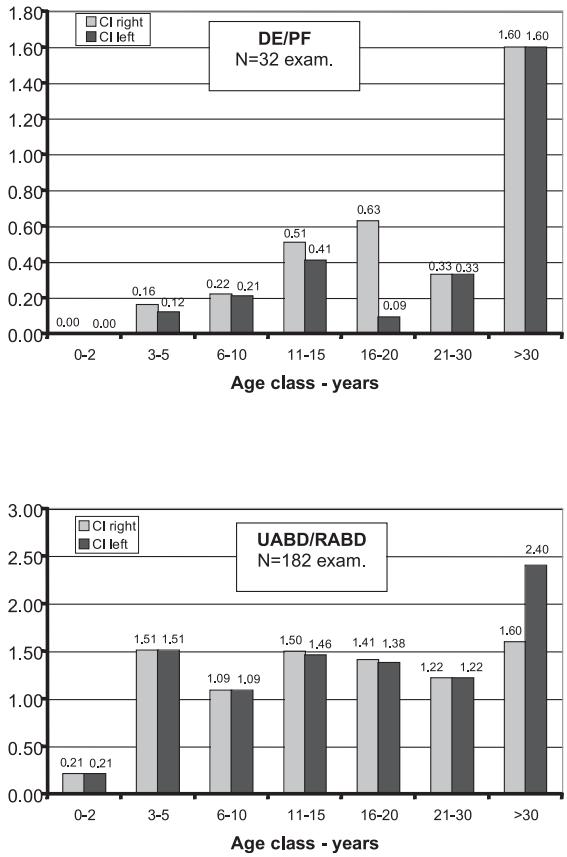

Fig. 4. Loss of range of motion (ROM) – relative contracture index (CI) – wrist joint DE/PF and UABD/RABD in SMA Type II

UABD/RABD were restricted in 182 cases (43.2%) and the CI remained relatively constant in all the age groups. Hand preference was not discernible from the data (Table 5 and Diagram 4). In the longitudinal data there was a noticeable and consistent decrease in RABD over time while UABD seemed to increase slightly. Ulnar abduction contractures were detected in 12 patients from various age groups (Table 6 and Diagram 5).

DISCUSSION

Contractures of the upper extremities are common in children with SMA II, and progress in frequency and severity over childhood. We have been unable to find any published papers with comparable numbers of patients and documentation to this study. Altogether there are very few detailed publications on this topic [5-10]. So far this has been the most extensive study of mobility and contracture development in the upper extremities of patients with SMA type II.

Because we divided the patients into age groups and because of the number of examinations, we were able to make more precise statements about those aged about 3 to 15. Younger and older patients had been examined fewer times so we could estimate only

Tab. 5. Loss of range of motion (ROM) of wrist joint in SMA Type II. Combined data for right and left limbs

Tab. 6. Manifest UABD-contracture of wrist joint in SMA Type II. Combined data for right and left limbs

Patients' age (years)	Number of examinations	Contracture present (%)	No contracture (%)	Manifest UABD-contracture [°]		
				Median	Mean (s_D)	Min - Max
0 - 2	12	0 (0%)	12 (100%)	-	-	-
3 - 5	71	0 (0%)	71 (100%)	-	-	-
6 - 10	203	4 (2%)	199 (98%)	7°	9.2° (5.8°)	5° - 20°
11 - 15	105	5 (4.8%)	100 (95.2%)	10°	15° (10.4°)	5° - 30°
16 - 20	16	1 (6.2%)	15 (93.7%)		5°	
21 - 30	9	0 (0%)	9 (100%)	-	-	-
> 30	5	2 (40%)	3 (60%)		30° and 30°	
All examinations	421	12 (2.8%)	409 (97.1%)	10°	14.1° (10.2°)	5° - 30°

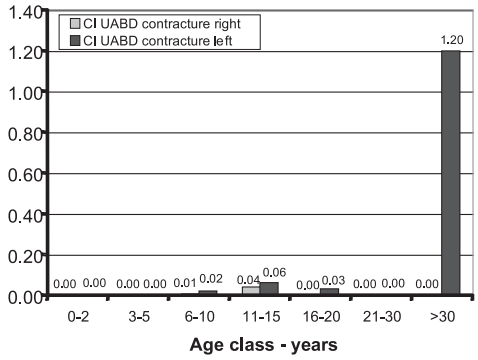

Fig. 5. UABD-contracture of wrist joint – relative contracture index (CI) – in SMA Type II. N=12 exam

roughly how the contractures would progress. Our observations showed a continual increase in the loss of range of motion and in contractures of the joints of the upper extremities as the patients got older.

This study includes more than one evaluation from each patient and a few patients can dominate the take-home line.

If one patient was evaluated several times during a particular age period and he manifested only the abnormality at the last evaluation, this could influence the data as the probability of recording an abnormal finding in that patient was higher compared to the individual who was seen only once at the beginning of the period. The choice for our evaluation of data from examinations with the greatest loss of range of motion could lead to subject selection, which would tend to magnify the effect of error and tend to bias the result toward greater impairment.

The combination of longitudinal and cross-sectional data in our study means that we are limited in our prediction of the actual individual potential for the development of contractures.

Restrictions of motion for all three ranges of shoulder movements were found, starting already from the ages of 3 to 5 years, with abduction/adduction mostly affected first. Rotation and flexion/extension were rarely restricted.

Johnson et al. examined 20 patients with chronic SMA at an average age of 37.9 years (1-75) and found a mean maximum loss of range of motion of 61° for shoulder abduction [5]. This corresponds with Wang et al.'s result of 51° [6]. In our study the average restriction of abduction/adduction was 6.9°. We noted two patients with abduction contractures but no adduction contractures.

Johnson et al. report that 32% of their patients had shoulder contractures [5]. Their definition of a contracture is a loss of range of motion of 5° [5]. This corresponds more or less to our percentage of patients of 38% and to 30% in Wang et al.'s study [6].

Wang et al. examined 27 patients with SMA type II aged 0.8 to 22.2 years (mean age 9.8±6.5) [6]. They found the following restrictions: restriction of shoulder flexion, shoulder abduction and elbow extension in 8 patients (30%), restriction of forearm supination in 7 patients (26%), and restriction of shoulder internal rotation, elbow flexion and wrist extension in 2 patients (7%). The greatest average loss of the range of movement was observed in this study with regard to forearm supination (70°), shoulder abduction (51°) and wrist extension (48°). Restriction of shoulder internal rotation was found only in older patients, above 15 years. No significant difference between the mobility of the joints of the right and left extremities was found but there was a positive correlation between restriction of mobility, increasing age and the functional status of the upper extremities [6].

Since the patient groups and methods are different, it is difficult to make a direct comparison of our CI values with those of Johnson et al. and Wang et al. [5,6]. Johnson et al. obtained the highest CI values for abduction of the shoulder and Wang et al. for forearm supination and shoulder abduction [5,6]. Our highest CI value was for the restriction of elbow extension, which indicates that the most marked restriction of motion in the upper extremities of patients with SMA type II appears in the elbow joint with, at times, massive flexion contractures.

The extent of the flexion contracture in the elbow increased steadily from the age of 3-5 (Table 3). The mean maximum loss of range of motion was about 40° in both our study and that of Johnson et al. and about 33° according to Wang et al. [5,6].

The mean loss of range of PRO/SUP was 55.1° in our study and 70° in Wang et al.'s study [6]. Loss of range of supination increased with age and was found in 32% of the patients in our group and in 26% of Wang et al.'s patients.

The mean reduction in the range of motion of DE/PF of the wrist was 60° according to Johnson et al., 40° in Wang et al.'s study and 42° in our patients [5,6]. Characteristic of SMA type II is early development of restricted radial abduction of the wrist, leading to ulnar abduction contractures in some patients (Tables 5 and 6).

Benady collected clinical findings of 50 children with intermediate and mild forms of SMA (SMA type II and III), reporting about 3 children with elbow flexion contractures and one child with a contracture of the long finger flexors [7]. There were mostly small children in this study, their ages at the last examination ranging between 13 months and 18 years.

After an average of 15 years' observation following diagnosis of SMA type II, Evans et al. found that 3 of their 21 patients had elbow flexion contractures of more than 40° [8]. They describe contractures of the upper extremities in SMA as being altogether rare and establish that the elbow is affected most and the wrist less often [8].

Willig et al. write about a group of 58 patients with SMA type II at a mean age of 12.6 years (±10.4) [9]. 16 had elbow flexion contractures of less than 25°, 12 had contractures exceeding 25° and 30 had no contractures. The functional restrictions that were reported in this study correlated with the severity of the contracture. Patients with elbow joint contractures more often reported elbow pain and sometimes shoulder pain [9].

Carter et al. examined joint mobility in 18 patients with SMA type II at a mean age of 17 years (±14) [10]. They found a restriction in mobility of at least 20° in the elbow of 22% of the patients and in the wrist of 44% of the patients.

To date there has been no scientific information about conservative (e.g. physiotherapy, orthoses) vs. surgical treatment of contractures of the upper extremities in patients with SMA. Further studies on this subject are needed.

CONCLUSIONS

1. This study shows the development of restrictions of passive range of motion in the joints of the upper extremities across the age groups in SMA type II patients.
2. Loss of range of motion and contractures of the joints of the upper extremities (shoulder, elbow and wrist) progressively increased with age.
3. The greatest restriction of motion was in the elbow joint with flexion contractures and loss of range of supination.

4. The results of this study may be used as a basis for comparisons in further studies dealing with the effectiveness of treatments to address both the contracture itself as well as the underlying cause of the contracture in SMA.

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