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Title:

Criterion validity of the Spatial Exploration Test of Upper Limb Mobility to evaluate the active horizontal workspace of children with spinal muscular atrophy.

Abstract

Purpose. To determine the criterion validity of the SET-ULM (Spatial Exploration Test of Upper Limb Mobility), a functional workspace test.

Materials and Methods. A prospective study from July 2017 to November 2018 in 30 children with SMA type 1 or 2. All children underwent assessment with the SET-ULM and the Motor Function Measure (MFM).

Results. We included 30 children. Median (Q1; Q3) MFM D1 (standing ability, ambulation and transfers), D2 (axial and proximal motor function), D3 (distal motor function) scores, Total MFM and Total SET-ULM active score were respectively 2.6% (2.6-3.8); 45.8% (19.9-65.3); 57.7% (36.9-80.9); 35.4% (16.7-43.2) and 70.2% (49.7-97.9). Total SET-ULM active score was strongly correlated with the MFM D2 dimension score (ρ 0.82; $p < 0.001$), with the D3 dimension (ρ 0.86; $p < 0.001$) and with the Total MFM score (ρ 0.89; $p < 0.005$). Total SET-ULM active score differed between SMA types ($p < 0.01$).

Conclusion. The SET-ULM has good criterion validity for the evaluation of available horizontal active upper limb workspace in children with SMA1 and SMA2. Future studies should evaluate reliability and sensitivity to change during a longitudinal follow-up study, as well as in a longitudinal trial of therapeutic effectiveness.

Clinical Trials: NCT03223051

Keywords: Spinal muscular atrophy • Children • Functional assessment • Spatial exploration • Upper limb

Introduction

Spinal muscular atrophy (SMA) is a severe, genetic, neurodegenerative disorder characterized by progressive muscle atrophy and weakness that first affects the lower extremities and then the trunk and upper limbs. The respiratory muscles and bulbar functions are affected in the most severe forms [1].

SMA spans a continuum of phenotypes, ranging from mild forms that are diagnosed in adulthood to severe forms with prenatal onset. Before 2016, no specific therapies were available to treat SMA. Affected children were classified as having SMA1, SMA2, or SMA3 depending on their age at the time of onset and the highest motor milestones achieved [1,4]. This classification is still the gold standard in clinical practice. Thanks to the development of several innovative treatments in the last decade and their availability in France (intrathecal SPINRAZA ® since 2017, intravenous ZOLGENSMA ® since 2019 and oral EVRYSDI ® since 2020), the natural course of the disease has changed considerably. Survival rates increase and motor function are improved when treatment is administered early, in particular for the most severe forms [4,7-16]. As a result of these major changes in prognosis, consensus groups [8, 9] have recently modified the classification by adding functional status (non-sitters, sitters and walkers) and the genetic background (number of *SMN2* copies).

All individuals with SMA1 and most of those with SMA2 have severe lower limb paralysis and variable limitations of upper limb function. Most have difficulty reaching objects placed in front of them. In contrast, the upper limb function of those with SMA3 is relatively spared until they lose walking ability.

The advent of treatments has led to a need for clinical tools to measure changes in upper limb function. Over the past 10 years, a variety of functional scales have been validated for use with children with SMA and are recommended by national and international consensus (PNDS <http://www.has-sante.fr>) [5, 10]: the CHOP INTEND [10], the Hammersmith Functional Motor Scale Expanded (HFMSE) [12, 13] [14, 15], Myotools [15], the Upper Limb Module (ULM) [16], the revised ULM (RULM) [16] and the Motor Function Measure (MFM) [17]. The HFMSE and MFM [18] are most frequently used in clinical trials but only the MFM and the RULM include items that assess upper limb function and dexterity [17, 18] [27-29]. The MFM is validated for use in children > 3 years up to adults in their 60s and covers a large spectrum of severities from no head control to ambulatory. The MFM differentiates between 3 types of motor function: standing and transfers (D1), axial and proximal limb motor function (D2) and distal upper motor function (D3). It is available in 2 versions, a version with 20 items for

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3 children aged between 2 and 6 years (MFM-20) and a version with 12 additional items for those > 6
4 years (MFM-32). However, the version for younger children lacks most items that assess upper limb
5 function in MFM32. The MFM is not sensitive to changes in movement speed or the ease with which
6 a task is performed. However, children and their caregivers often describe subtle but clinically
7 meaningful functional improvements after the introduction of a therapy that are not detected by
8 currently available motor scales. This is particularly true for horizontal upper limb movements, i.e.,
9 not against gravity, like reaching, colouring, and pointing, and that can be improved with assistive
10 devices (e.g., suspended arm supports). Furthermore, no current tools evaluate supported arm function
11 in young children.
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20 To fill this gap, we developed a new evaluation tool, the Spatial Exploration Test for Upper Limb
21 Mobility (SET-ULM). The SET-ULM assesses the dimension active workspace in the horizontal
22 plane. It was designed to provide complementary information to the MFM scale and to be feasible for
23 use in young children with marked arm weakness. The SET-ULM measures active reaching workspace
24 and might therefore capture small changes in upper limb motor function in the horizontal plane.
25 The primary aim of this study was to determine the criterion validity of the SET-ULM. Criterion
26 validity is an estimate of the extent to which a measure agrees with a gold standard, here the MFM.
27 We hypothesised that SET-ULM and MFM scores would be strongly correlated. The secondary aims
28 were to determine if the SET-ULM could differentiate between children with SMA1 and SMA2 and
29 between different age groups (3-9 years vs 10-16 years: according to the World Health Organisation,
30 the maturation of central nervous system is more marked between 3 to 9 years than between 10 to 16
31 years).
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41 **Methods**

42 *Participants*

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44 Children were recruited from the Neuromuscular Unit of Raymond Poincaré University Hospital,
45 Garches, France. Inclusion criteria were: aged between 3 and 16 years, genetic confirmation of SMA,
46 inability to walk, and parental consent for participation. Exclusion criteria were any cognitive
47 dysfunction or the presence of another pathology that would prevent performance of the test. The study
48 was approved by the Lyon III Ethics Committee (2017-A01017-46). Parents provided written informed
49 consent and the children were given an information letter appropriate for their age.
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58 *Materials Methods*

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3 We conducted a 16-month, single centre, interventional, prospective trial (ExplorASI: 2D workspace
4 exploration in children with Spinal Muscular Atrophy). Each participant underwent a single evaluation
5 that included the MFM and the SET-ULM. The order of the tests was randomized to avoid any bias
6 caused by fatigue. All the assessments were performed by the same occupational therapist.
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10 11 *MFM*

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15 The MFM assesses standing ability, ambulation and transfers (D1 sub-score), axial and proximal motor
16 function (D2 sub-score), and distal motor function (D3 sub-score). Items are scored on a 4-point Likert
17 scale based on the individual's maximal ability without assistance: 0: does not initiate movement or
18 starting position cannot be maintained; 1: partially completes the exercise; 2: completes the exercise
19 with compensations, slowness or obvious clumsiness; and 3: completes the exercise with a standard
20 pattern. To enable comparison with other tests and because 2 different MFMs were used to
21 accommodate the different ages of the children, the results are expressed as a percentage of the
22 maximum possible score.
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28 29 *SET -ULM*

30 31 32 *SET-ULM Material*

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35 The SET-ULM was designed and developed in our lab to evaluate active workspace (active reaching
36 distance over the whole horizontal plane).
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39 ----- Insert figure 1-----

40 The test involves pointing to targets on a test board 170 cm wide and 60 cm deep (Figure 1). The board
41 and targets were designed to be child-friendly with targets that were easy to recognise by children to
42 avoid any misunderstandings. Two groups of targets were drawn on the board: 11 far targets (purple)
43 were positioned according to anthropometric data for healthy 16-year-olds and 11 close targets (red)
44 were positioned according to data for healthy 3-year-olds. We used (median) anthropometric data
45 recorded in each child's health book received at birth for their medical follow-up. These limits were
46 determined with the trunk constrained since the majority of individuals with SMA who might perform
47 the test wear a Garches brace with a pre-sternal breast piece, and head and chin support (Figure 2). A
48 yellow sun was positioned in the centre at the bottom of the board and was the starting position for all
49 the movements.
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7 *SET-ULM Assessment*
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10 The SET-ULM was performed in a standardized position: the child was seated comfortably in a chair
11 or wheelchair with their feet supported so that their ankles, knees and hips were flexed at 90° (on the
12 floor, on a step or on the footrests of the wheelchair). Their abdomen (the brace) touched the edge of
13 a height-adjustable table, their elbows were flexed at 90°, shoulders relaxed. In addition, the trunk was
14 fixed with a belt to avoid any compensatory flexion, lateral flexion or rotation movements.
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20 The evaluator first asked the child to name each target to ensure that they would understand the
21 instructions and checked that the brace did not block their view of any targets. The evaluator then
22 passively extended and moved each of the child's upper limbs in turn through horizontal abduction
23 and adduction and placed 11 stickers on the arc formed by the child's extended index finger; yellow
24 stickers (8 mm diameter) were used for the right limb and green for the left (Figure 3). The evaluator
25 then asked the child to attempt to point to each of the stickers in turn (actively) as close as possible to
26 the point reached passively. The child began with their dominant or preferred arm. The child could use
27 compensatory movements (eg. "crawling" with the fingers over the board). Before each exercise, the
28 examiner placed the child's index finger on the starting target (the yellow sun, Figure 1). The evaluator
29 provided encouragement throughout the test and placed a sticker at the furthest point reached by the
30 index finger for each movement (blue sticker for the left limb and red for the right).
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39 Two curves were then drawn for each limb by connecting the points to determine the passive and active
40 horizontal workspaces (Figure 3).
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45 *Measures*
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50 *SET-ULM Scores*
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53 The coordinates (x, y) of each point for the active and passive workspaces were measured using a
54 measuring tape and then entered into an Excel file established for the calculation of the different scores.
55 The areas of each of the 3 workspaces, theoretical, passive and active, were determined by calculating
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3 the area under the curve. We used the Conventional Trapezoid method; the total area is decomposed
4 into sub areas. Trapezoids are formed by linear interpolation between each point.

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7 Three SET-ULM scores, expressed as percentages, were calculated: a theoretical score and passive
8 and active scores.

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10 The theoretical score is the ratio between the active area and the theoretical area; the theoretical area
11 represents the area explored by a healthy subject with equal arm size, reported as a percentage.

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13 The **passive SET-ULM score** is the ratio of the passive area to the theoretical area and represents the
14 area available to the child taking joint limitations or other upper limb orthopaedic deformities into
15 account, reported as a percentage.

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18 The **active SET-ULM score** is the ratio of the active area to the passive area and represents the area
19 that actively available to the child according to the length of his or her upper limbs (obtained passively),
20 reported as a percentage. Separate active scores are calculated for the **left** (green and blue stickers) and
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23 **right** limbs (yellow and red stickers).

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26 The **total SET-ULM active score** (area under the curves) is calculated from the two maximum curves
27 obtained by connecting the 11 furthest points. It is not the sum of the two areas since that would involve
28 superimposing the common surface explored by the two upper limbs which corresponds to the area for
29 bimanual functional activities.

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32 The curves on the test board provide a representation (cartography) with 2 results, the passive zone,
33 and the actual zone reached by each child in the horizontal plane. The active curve provides a quick
34 visual representation of muscle weakness while the passive curve highlights the presence and severity
35 of joint limitations or upper limb orthopaedic deformities.

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38 The passive score is useful for comparison of the child's movement with that of a healthy subject.
39 However, the particular interest of this test is to quantify active function. We therefore focused the
40 analysis on the active score that reflects the active horizontal workspace available to the children with
41 SMA, named "SET-ULM Score".

42 43 44 45 46 47 48 *Data analysis*

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51 We calculated medians and (Q1; Q3) for continuous variables and frequencies for categorical variables.
52 The data did not have a normal distribution (Shapiro-Wilks's test) therefore we used non-parametric
53 tests. A Spearman correlation with Bonferroni correction was used to analyse the relationship between
54 the total SET-ULM active score and MFM D1, MFM D2, MFM D3 scores, and MFM total score.
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We compared total SET-ULM active scores between the SMA1 and SMA2 groups, and between the 3–9-year-old group and the 10–16-year-old group with a Wilcoxon test. We used a Spearman correlation with Bonferroni correction to determine the relationship between the left and right SET-ULM active scores and between treated/untreated children. In addition, we assessed the correlation of the total SET-ULM active scores between the SMA1 and SMA2 groups treated and untreated. The level of significance was fixed at $p < 0.01$. All statistical analyses were performed using R Studio® Version 3.3.3 software.

RESULTS

Participants

Of 80 eligible participants, 30 children aged 3 to 16 years agreed to participate and completed the assessment.

Participant characteristics are shown in Table 1.

----- Insert table1 -----

Total SET-ULM active scores and MFM scores

Table 2 shows D1, D2, D3 and total MFM score, and Total SET-ULM active score for each participant.

----- Insert Table 2 -----

Strong correlations were found between total SET-ULM active score and D2 (axial and proximal motor function) ($\rho = 0.82$, $p < 0.01$), total SET-ULM active score and D3 (distal motor function) ($\rho = 0.86$, $p < 0.01$) and total SET-ULM active score and MFM total score ($\rho = 0.89$, $p < 0.01$). Weak correlations were found between total SET-ULM active score and MFM D1 "standing and transfers" ($\rho = 0.43$, $p = 0.02$).

Total SET-ULM active score by SMA type and age.

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3 Table 3 shows the total SET-ULM active score for each SMA type (type 1 and type 2) and each age
4 group (3-9 years and 10-16 years).
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10 The median total SET-ULM active score for all participants was 70.25 % (Q1 49.7; Q3 97.9). There
11 was a significant effect of the type of SMA on total SET-ULM active score ($p < 0.01$) but no effect of
12 age ($p = 0.40$).
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19 *Total SET-ULM right and left active scores for all participants*
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21 Total SET-ULM active right and left scores were strongly correlated ($\rho = 0.92$, $p < 0.01$), showing
22 overall symmetrical performance.
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27 *Total SET-ULM active score by treatment group (treated/untreated).*
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29 Total SET-ULM active score was strongly correlated ($\rho = 0.92$, $p < 0.01$) with MFM total score in
30 untreated children ($n = 21$) but not ($\rho = 0.53$, $p < 0.14$) treated children ($n = 9$).
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35 **Discussion**
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38 This study demonstrated that the Spatial Exploration Test (SET) of Upper Limb Movement (ULM) is
39 a pertinent new tool to assess the active horizontal workspace of children with SMA 1 and 2 aged 3 to
40 16 years. Furthermore, the strong correlations between the SET-ULM and MFM scores, likely suggest
41 the test has good criterion validity.
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46 To our knowledge, this is the first test designed to map the active horizontal workspace of individuals
47 with neuromuscular disease, in particular children with SMA. The 3 active scores obtained from the
48 test, (SET-ULM right, SET-ULM left and total SET-ULM), expressed as percentages of the
49 individual's passive score, correspond to the surface actually explored by the child in each zone,
50 relative to the length of their upper limbs.
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54 We consider that this tool is relevant for the purpose for which it was created since correlations
55 between the Total SET-ULM active score and the MFM scores demonstrated strong criterion validity.
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3 Correlation with the distal dimension (D3) scores was slightly stronger than with the axial and
4 proximal dimension (D2) scores and the total MFM scores. This is not surprising since D2 involves
5 evaluation of functions such as head control, which are not included in the SET-ULM, and the total
6 score included D1, which involves walking ability. The SET-ULM provides precise information
7 regarding the amplitude that the child can reach actively in the horizontal plane and is thus
8 complementary to the MFM, which provides an indication of the child's level of function. The strong
9 correlation between the D3 score and the Total SET-ULM score could also be explained by the fact
10 that movements such as "crawling" with the fingers over the board, which are not considered in
11 functional tests, can be used in the SET-ULM to reach the target. This further demonstrates that the
12 SET-ULM is complementary to the MFM. Importantly, the SET-ULM was easy to use and the time
13 to measure and process the results was around 15 minutes (including the SET-ULM right, SET-ULM
14 left, total SET-ULM, measurements of area under the curve, and the visual graphical representation of
15 the child's passive and active workspace).

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26 The SET-ULM successfully differentiated between the children with SMA1 and SMA2 since the
27 scores of the children with SMA1, who have more severe muscle weakness, were significantly lower
28 than those of the children with SMA2, who have a higher functional capacity of the upper limbs. In
29 contrast, and contrary to our expectation, the results were not affected by age group, which suggests
30 that the SET-ULM is adapted and pertinent for the measurement of workspace throughout childhood
31 and whatever the type of SMA (1 or 2).

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36 There was also a strong correlation between the results of the right and left SET-ULM scores. This
37 was expected and corresponds to the symmetrical motor impairment caused by SMA described in the
38 literature [21]. However, in clinical practice it is not uncommon to find individuals with a degree of
39 asymmetrical involvement. In contrast with the MFM, in which the child performs the task with only
40 their preferred hand, the SET-ULM can measure interlimb asymmetries caused by different degrees of
41 weakness or asymmetrical orthopaedic deformities. This is another aspect in which the SET-ULM is
42 complementary to the MFM.

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48 MFM total and SET-ULM active scores were not correlated in the treated children. However, sample
49 bias may be present (9 versus 21 children). In addition, we did not consider the dates of treatment
50 administration in relation to the timing of the evaluations for the study. We cannot affirm, given this
51 information and the lack of hindsight on these new treatments, that the treatment effects were still
52 optimal at the time of the SET-ULM evaluation. The sensitivity and specificity of the SET-ULM
53 remain unknown. These metrics should be determined before carrying out longitudinal follow-up of
54 individuals with SMA to evaluate treatment effects.

Limitations

The main limitation of this study is that the number of children included was relatively small and they were heterogeneous in terms of age and clinical baseline characteristics, however this reflects the real-life situation of children with SMA in France today.

Conclusion

This study demonstrated the criterion validity of the SET-ULM. This test is a useful adjunct to the MFM since it provides a precise evaluation of horizontal reaching ability. This study provides pilot data for a new test that may be clinically useful once reliability and sensitivity have been established during a longitudinal follow-up study, as well as in a longitudinal trial of treatment effectiveness. This test has many perspectives. In this study, we only evaluated pointing and did not include grasping, which is a highly important aspect of upper limb functional capacity. The test could easily be developed to include a measurement of grasping workspace area. One of the strengths of the SET-ULM in its current form is its low dependence on technology, potentially allowing for widespread cost-effective uptake. However, it could also be combined with 3D motion analysis or accelerometers to evaluate upper limb kinematics. Such an analysis could be used to determine exploratory strategies used by individuals with SMA during a reaching or a grasping task and if these strategies depend on the type of SMA or change with treatment. Although we focused on children with SMA because of the lack of evaluation tools in this population, the SET-ULM could be used to evaluate upper limb function in different neuro-muscular diseases that affect upper limb movement.

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Declaration of interest.

The authors report there are no competing interests to declare.

Data Statement

The datasets generated and analysed during the current study are available from the corresponding author on reasonable request.

Statement Of Ethics

The study was approved by the Lyon III People's Protection Committee

Author Contributions

Sandrine Pruvost, Samuel Pouplin and Nicolas Roche contributed to the development of the SET-ULM board

All the authors contributed to the writing of the manuscript and read and approved it

The experiments were performed by Sandrine Pruvost

Legends

Figure 1: the SET-ULM Board. In red, targets that correspond to anthropometric data for healthy 3-year-olds, in purple, targets that correspond to anthropometric data for healthy 16-year-olds. The yellow sun in the bottom centre was the starting position for all movements. The board was designed to be child friendly.

Figure 1 Alt Text: the SET-ULM Board is presented here. It consists of a board showing different targets. In red, targets that correspond to anthropometric data for healthy 3-year-olds, in purple, targets that correspond to anthropometric data for healthy 16-year-olds. The yellow sun in the bottom centre was the starting position for all movements. The board was designed to be child friendly.

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3 Figure 2: a child wearing a Garches brace with a pre-sternal breast piece, and head and chin support,
4 performing the SET-ULM assessment.
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7 Figure 2 Alt Text: the figure presented a young child wearing a Garches brace with a pre-sternal
8 breast piece, and head and chin support, performing the SET-ULM assessment
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11 Figure 3: Examples for two children: the position of the passive targets for the left and right arms are
12 shown in green and yellow respectively and the targets reached actively are shown in blue and red.
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14 The scores are shown under the figure.
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16 Figure 3 Alt Text: Two children assessment are presented here. The position of the passive targets for
17 the left and right arms are shown in green and yellow respectively and the targets reached actively are
18 shown in blue and red. The scores are shown under the figure.
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23 Figure 4: Trial flow chart.
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25 Figure 4 Alt Text: this is the flow chart of the study. This diagram shows the inclusion and non-
26 inclusion criteria, the assessments performed, the people lost to follow-up and the number of people
27 analyzed.
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31 Table 1: Demographic description of participants
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35 Table 2: D1 (standing and transfers), D2 (axial and proximal limb motor function), D3 (distal motor
36 function) scores and total MFM (Motor Function Measurement) and Total SET-ULM (Spatial
37 Exploration Test for Upper Limb Mobility) active score (%).
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41 Table 3: Total SET-ULM active score for each SMA type (type 1 and type 2) and each age group (3-
42 9 years and 10-16 years)
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Title:

Criterion validity of the Spatial Exploration Test of Upper Limb Mobility to evaluate the active horizontal workspace of children with spinal muscular atrophy.

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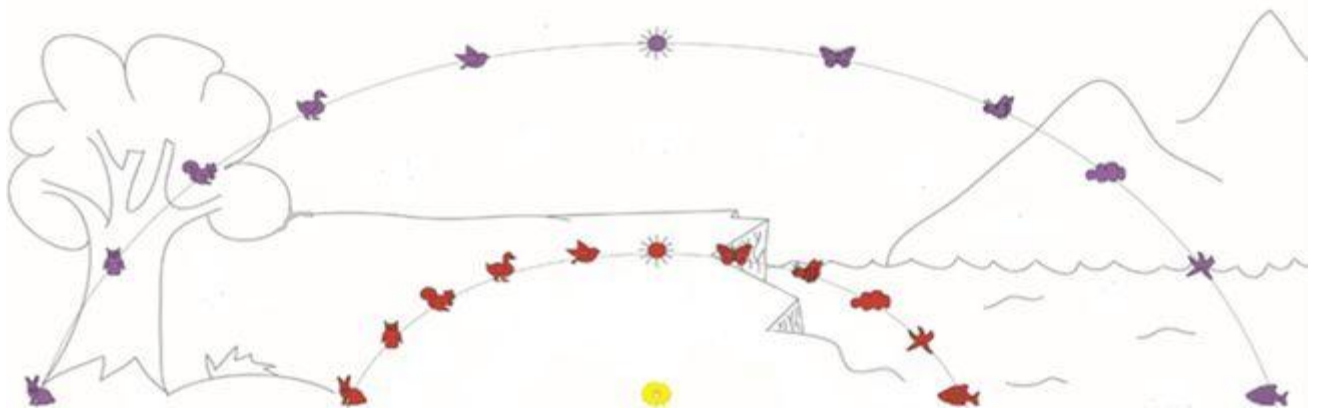
Number of references: 22

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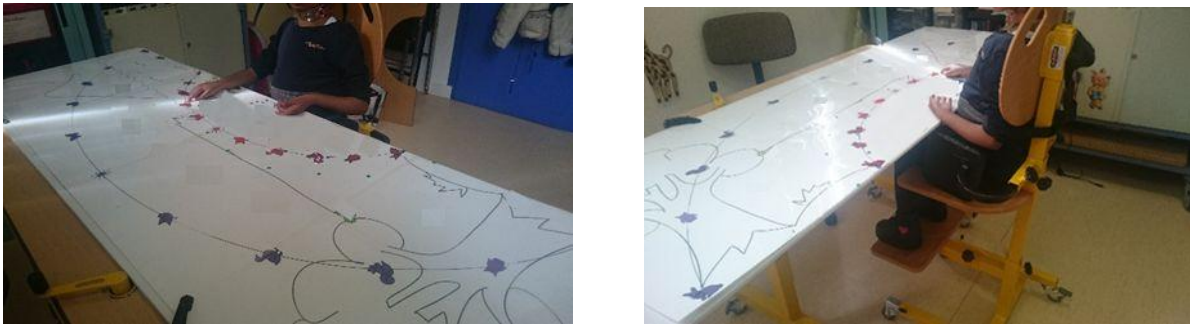
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3 Figure 1: the SET-ULM Board. In red, targets that correspond to anthropometric data for
4 healthy 3-year-olds, in purple, targets that correspond to anthropometric data for healthy 16-
5 year-olds. The yellow sun in the bottom centre was the starting position for all movements. The
6 board was designed to be child friendly.
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27 Alt Text: the SET-ULM Board is presented here. It consists of a board showing different targets.

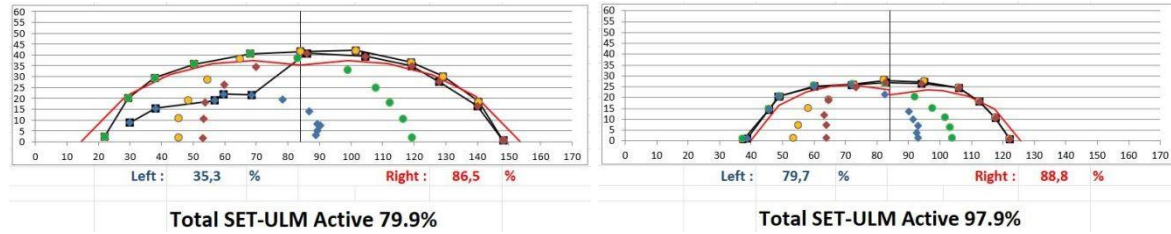
28
29 In red, targets that correspond to anthropometric data for healthy 3-year-olds, in purple, targets
30 that correspond to anthropometric data for healthy 16-year-olds. The yellow sun at the bottom
31 centre was the starting position for all movements. The board was designed to be child friendly.
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3 Figure 2: a child wearing a Garches brace with a pre-sternal breast piece, and head and chin
4 support, performing the SET-ULM assessment.
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21 Alt Text: the figure presents a young child wearing a Garches brace with a pre-sternal breast
22 piece, and head and chin support, performing the SET-ULM assessment
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Figure 3: Examples for two children: the position of the passive targets for the left and right arms are shown in green and yellow respectively and the targets reached actively are shown in blue and red. The scores are shown under the figure.



Alt Text: Assessments for two children are presented here. The position of the passive targets for the left and right arms are shown in green and yellow respectively and the targets reached actively are shown in blue and red. The scores are shown under the figure.

Table 1: Demographic description of participants

			N	Age (years) median (Q1; Q3)
Participants	<i>All</i>		30	10.0 (8.0; 11.7)
Sex	<i>Male</i>		15	9.0 (8.0; 11.0]
	<i>Female</i>		15	10.0 (8.5; 12.0)
SMA Type	<i>SMA1</i>	<i>Total</i>	8	10.5 (8.5; 11.7)
		<i>(3-9 years)</i>	3	7.0 (5.5; 8.0)
		<i>(10-16 years)</i>	5	11.0 (11.0; 14.0]
	<i>SMA2</i>	<i>Total</i>	22	9.5 (8.0; 11.7)
		<i>(3-9 years)</i>	11	8.0 (7.5; 9.0)
		<i>(10-16 years)</i>	11	12.0 (10.5; 13.0)
Laterality	<i>Right-Handed</i>		21	10.0 (9.0; 12.0)
	<i>Left-Handed</i>		5	9.0 (7.0; 14.0)
	<i>Ambidextrous</i>		3	9.0 (8.5; 10.0)
	<i>Not acquired</i>		1	4.0
Treatment (Nusinersen)	<i>SMA1</i>	<i>Yes</i>	5	11.0 (7.0; 11.0)
		<i>No</i>	3	10.0 (9.5; 12.0)
	<i>SMA2</i>	<i>Yes</i>	4	9 (8.7; 9.2)
		<i>No</i>	18	10 (8.0; 12.0)

Table 2: D1 (standing and transfers), D2 (axial and proximal limb motor function) and D3 (distal motor function) scores, and total MFM (Motor Function Measure) and Total SET-ULM (Spatial Exploration Test for Upper Limb Mobility) active score (%).

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Participant	MFM D1 (%)	MFM D2 (%)	MFM D3 (%)	MFM Total (%)	SET-ULM Total Active (%)	Area under the curve in cm ²
1	2.6	44.4	90.5	37.5	91.2	3538
2	2.6	61.1	80.9	41.7	91.5	3400
3	2.6	69.4	80.9	44.9	99.5	2661
4	2.6	11.1	33.3	12.5	22.3	364
5	2.6	8.3	4.8	5.2	7.8	166
6	10.6	94.4	100.0	61.5	100.0	4123
7	5.1	72.2	80.9	46.9	99.3	2361
8	5.1	80.5	80.9	50.0	97.9	2820
9	0.0	5.5	28.6	8.3	8.3	200
10	0.0	52.8	71.4	35.4	79.9	3308
11	5.1	94.4	90.5	57.3	100.0	4498
12	2.6	41.7	57.1	29.2	45.8	1476
13	4.7	66.7	58.3	40.0	97.9	1795
14	2.6	72.2	71.4	43.7	64.3	1786
15	2.6	38.9	52.4	36.5	79.5	2205
16	0.0	5.5	9.5	4.1	0.7	27
17	5.1	58.3	90.5	43.7	100.0	3460
18	0.0	41.7	47.6	26.0	62.1	2352
19	4.2	20.8	50.0	20.0	54.1	1206
20	0.0	38.9	100.0	36.5	99.3	3832
21	2.6	47.2	47.6	29.2	82.8	1922
22	2.6	19.4	33.3	15.6	19.4	278
23	2.6	72.2	95.2	49.0	100.0	5119
24	2.6	36.1	57.1	27.1	50.3	2255
25	4.2	16.7	33.3	15.0	53.6	1040
26	2.6	50.0	71.4	35.4	49.5	2943
27	0.0	50.0	66.7	33.3	74.5	2344
28	2.6	2.8	14.3	5.2	0.0	0
28	2.6	13.9	28.6	12.5	66.0	2268
30	2.6	58.3	57.1	35.4	60.9	1389
Median	2.6	45.8	57.7	35.4	70.2	2261.5
(Q1 ; Q3)	(2.6 ; 3.8)	(19.8 ; 65.3)	(36.9 ; 80.9)	(16.7 ; 43.2)	(49.7 ; 97.9)	(1251.7 ; 3216.7)

Table 3: Total SET-ULM active score for each SMA type (type 1 and type 2) and each age group (3-9 years and 10-16 years)

			Total SET-ULM active score % (median, Q1, Q3)
All participants			70.2 (49.7- 97.9)
SMA Type	<i>SMA1</i>	<i>Total</i>	20.8 (8.2-47.7)
		<i>(3-9 years)</i>	19.4 (13.6-36.5)
		<i>(10-16 years)</i>	22.3 (8.3-45.8)
	<i>SMA2</i>	<i>Total</i>	87.0 (62.6-99.0)
		<i>(3-9 years)</i>	79.5 (61.5-98.0)
		<i>(10-16 years)</i>	91.2 (77.20-99.6)

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3 **Implications for rehabilitation**
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- 7 • The Spatial Exploration Test for Upper Limb Mobility is a useful adjunct to the Motor
8 Function Measure.
- 9 • It provides a precise evaluation of horizontal reaching ability.
- 10 • The Spatial Exploration Test for Upper Limb Mobility will be of great clinical utility
11 for the evaluation of the effects of treatments for spinal muscular atrophy.
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