

Workshop report
1st Sitting Workshop
for neuromuscular disorders
Denmark, 30 September - 1 October 2019

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Received 26 February 2020

1. Introduction

From 30th September to 1st October 2019 a workshop was organized on sitting for persons with neuromuscular disorders, specifically focusing on Duchenne muscular dystrophy (DMD), Spinal muscular atrophy (SMA), congenital muscular dystrophy (CMD) and myopathy (CM), in Musholm Ferie-Sport-Konference in Korsør, Denmark. Recruitment for occupational therapists was done in Western European countries; participating countries were Denmark, Norway, Sweden, Belgium, France and the Netherlands. A pre-workshop questionnaire was sent around concerning the use of standard evaluating methods and possible interventions.

The objectives of the workshop were to reach consensus on assessment of sitting posture, consensus on possible solutions for posture abnormalities, and starting the development of international seating guidelines and starting sitting research.

1.1. Identifying sitting problems

Weakness in the trunk and pelvic muscles leads to aberrant sitting postures. Both (kypho)scoliosis formation and asymmetric contractures around the hips can lead to unbalanced sitting, meaning the head is not in line with the middle of the pelvis. Pain and fatigue are frequently mentioned problems [1,2] (presentation by Ulla Werlauff). Next to this, aberrant sitting postures influence the lungs, heart and abdomen [3–5] (presentation by Imelda de Groot). Also, recent research was presented on the interaction between arm activities and trunk in patients with DMD and SMA [6–8]. From this research it is clear that there is a

difference in trunk control between DMD and SMA. Thus stabilizing the trunk needs to be approached differently to maintain optimal arm functionality. Message: Don't just look at the trunk, look at the arms as well.

1.2. View of a patient on sitting evaluation

Mr. Simon Jespersen, a patient representative with SMA type 2, told about his experience in adapting sitting posture. He has been sitting all of his life; however now as an adult he understands why his sitting position is important. In his presentation he looked back at the days with the therapists who adjusted his sitting posture being seated on a table, with no support, and being touched all over his body. As a child he did not understand the background and necessity of the adjustment. He wanted to maintain his strength and function and being corrected to a more upright position in his wheelchair made him feel that he – for a period – lost function. Change of mindset came when function declined. It made him more motivated, and he realized that the work of the occupational therapist is important. If he had seen people with a severe scoliosis as an example as to what could happen if you do not correct a posture, that would have motivated him to take action.

Simon's advice: it's all about compromise; tell the child that the small disadvantages by correcting the seating position now, may give them a big advantage in the future. As advice he also gave: make use of role models.

From the following discussion it became clear that parents play a role in sitting evaluation and proposed interventions. Also for parents it is difficult to envisage the consequences for the future; sometimes it seems easier to convince the child than the parents.

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2. Practical approaches in different countries

The pre-workshop questionnaire revealed that the participants all use different assessment procedures. The same was the case for possible interventions, many different approaches were described.

In the workshop all participants presented their way of evaluating sitting posture and possible interventions. France and the Netherlands used forms with drawings to describe the sitting posture. Sitting evaluation takes time depending on the approach: 30 min to 2,5 h. Different interventions are used: abdominal belts that are electrically adjustable in length, soft bracing, supportive pads / cushions fixed at the wheelchair frame, or individual seating by means of a moulded seat.

Two patients were willing to be experimental subjects for evaluating sitting posture. Both patients were discussed among the workshop participants, while sitting in the wheelchair and while being examined on the bench. It showed that in evaluation many common elements are present. However, on intervention there were different ideas.

An important question was raised not to be answered as yet: how can we adapt our devices during the day? What makes the person stable in the morning may not work in the evening because they collapse during the day.

3. Conclusion of this first day and new goal

As there are common elements in evaluation, though not structured and documented in a uniform way, and as there is a wide variation of possible interventions, we decided to adjust the goal of the workshop. Before it is possible to evaluate the effectiveness of different interventions in research a common means of evaluation is necessary. Therefore, on the second day we adjusted our goal and focused on:

- Can we come to an agreement on assessment?
- Can we make a collaborative working group to set up an assessment and intervention scheme?

4. Common assessment of sitting position

This session was done in a general discussion with all participants in a semi-structured way with statements on slides moderated by Imelda de Groot, based on the results of the discussions of the first day.

It was agreed that the prerequisites for a standard assessment of sitting position are: not time consuming, easy to use with no interpretation difficulties, useful for many countries, leads to insight in the problems and leads to a conclusion.

In France the SPCMA [9] is used for evaluation: seated Postural Control Measure for Adult version 2.0. This was discussed for applicability and compared to the Dutch evaluation form, which has comparability, but adds three different ways of observation. The Dutch form is developed by the collaborating occupational and physiotherapists of

the Dutch patients' organization, Spierziekten Nederland, of which Yolanda van den Elzen is one of the leading persons [10].

The use of pressure assessments is not uniform between the participating countries both in the way it is performed and the therapeutic indications. Therefore it is not considered as being a standard evaluation method.

It was agreed to use the 3 step approach of the Dutch working group on seating in NMDs. This approach starts with the anamnesis and then an observation and examination in 3 steps: observation in the wheelchair of the sitting position, observation and examination supine on an examination bench, and if possible sitting on the examination bench to explore correcting posture possibilities.

The proposed anamnesis:

Basic information:

- Patient's description of the main problem(s)
- Medical history; additional diagnoses
- Who are the caregivers (important in connection with transfers)
- Medical history
- Weight/height/BMI
- Pressure sore(s)
- Pain: yes/no; type of pain: neuropathic or nociceptive; location; time; can it be influenced by medication or change of position
- Respiratory problems
- Swallowing problems, tube feeding, PEG
- Urinary problems

Daily activities:

- Communication - Speech/loudness of voice
- Writing/reading/telephone/computer and gaming
- How long time do you sit in the wheelchair
- Arm/hand function - Preferred hand
- Household
- School work
- Sports
- Leisure
- Hobbies

Wheelchair:

- Type of chair and how old
- Type of seat, type of cushion (how old)
- Type of back rest
- Type of neck rest
- Type and place of joystick – is it heavy to move/spring
- Use of electric function – e.g. to change position
- Individual adaptation
- What is already tried as adaptations, and what functioned or what not
- Environment/surroundings/type of housing
- How temperature affects the person

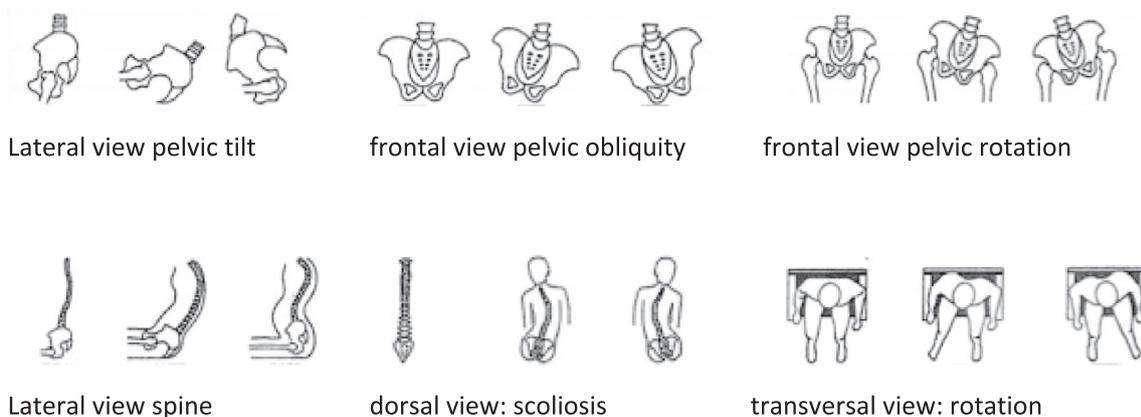


Fig. 1. Examples of the figures in the observation form.

The three forms of the Dutch seating workgroup were discussed, these forms contain figures which can be marked and give an easy overview. Adjustments, additions of figures and translation of these forms to English, and consequently in the language of each participating country, was agreed on (Fig. 1).

5. Interventions

5.1. Correcting sitting posture

After discussion the following was agreed on: concerning interventions for sitting posture it depends whether a position is rigid/fixe (not able to correct) or flexible (able to correct). In a fully rigid posture one must accept the position and balance the posture with supportive adjustments in the wheelchair. If the posture is still flexible, balance can be established with or without adjustments. In practice the participants agreed that it is neither rigid or flexible. The approach should be stepwise for both. Sometimes you do adaptations because of the severity of the problem. All participants agreed that there is always a compromise (related to function) – how much will be compromised related to function.

5.2. Use of braces

The use of braces was discussed. They are more frequently used in young children than in adults, although some adults prefer a brace. Especially in SMA, CMD and FSHD (when wheelchair confined) braces are used. Mostly this is for children with weak trunk muscles, defined as MRC grade 2–3 or hardly able to move against gravity, and if the child is not strong enough to hold up the trunk. When providing a child with a brace breathing/ventilation should be checked.

Different types of braces are used: in Gothenburg for children with SMA1 a hard/rigid brace is used like the Boston type (approximately each year a new one). They have no skin problems with skilled orthopedic engineers. In Denmark a T-brace is used, which leaves room for ribcage to allow for

respiration; for very small children with SMA type 1 a shield is used. In France they use a brace including a head support for children under two years of age, when they begin to sit until the surgery. Sometimes parents take off chin support, but this is not recommended. Therapists do not recommend the soft braces, as they never fit the child well enough, and do not adjust for scoliosis.

The use of an adjustable electrical belt was discussed as it is frequently used in Denmark and Norway, but not elsewhere. The Danish participants and patients elucidated the use of a belt. Two positions for use of electrical belt were mentioned: (1) if you have freedom to move back- and forward you have freedom to place the belt either more on the upper or the lumbar part of the trunk; (2) if the sitting position is with an extreme lordosis, the belt is placed around the stomach to support an upright position. In the latter one should be careful about the pressure on this area as it could influence digestion and gastrointestinal function. The patient can change position in the sagittal plane themselves (the belt can be loosened or fastened via the joystick). The advantage of this is that it makes it possible for the patient to relax and save strength for purposes other than sitting e.g. to use his arms.

The experience in the Netherlands is that sitting with arm supports also stabilizes the trunk, and one can also use a plate in front of the patient which allows him or her to lean against it, thus no need of supportive belt. New developments with soft exoskeletons are upcoming, as there are developments in several countries on this topic.

It became clear that no scientific evidence is available to make a weighted choice of interventions.

5.3. Overall conclusions

The need of a standardized screening method to evaluate sitting posture both seated in the wheelchair and not in the wheelchair was underlined by all participants. There was an agreement that the starting point is the Dutch form that needs to be adjusted based the discussion in this workshop.

There is no agreement on the type of interventions to adjust a sitting posture or to support a sitting posture. There is a lack of evidence in this field.

6. Next steps and creating a network

It was decided that first the draft evaluation form for a common standardized screening assessment should be tested among the participants of this workshop. This will be the topic of a next workshop to optimize the form and the assessment. A common assessment form can be the base of scientific studies on sitting interventions.

Next other countries will be asked to join the working group to use the standardized assessment and to collect data on interventions as used in their countries and the effects of these interventions after a specific time period. From this more studies can be started to evaluate efficacy of interventions and to optimize sitting posture and ability to perform daily activities for persons with neuromuscular disorders in a wheelchair.

Acknowledgments

This workshop was made possible by the Danish Muskelvindfonden and by the Dutch Duchenne Parents Project.

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