INNOWALK – BENEFICIAL EFFECT IN SPINAL MUSCULAR ATROPHY

By Ulrika Skjellvik Tollefsen

There are many exciting aids on the market nowadays. One is Innowalk, a motorised aid that provides assisted movement in a standing position for children with impaired mobility. It is so new on the market that there are no studies of its effect.

Several case studies have been carried out involving Innowalk, but to date no articles have been published. One study is in the process of publication and several others have been started, mostly concerning users with cerebral palsy (CP).

My name is Ulrika Skjellvik Tollefsen and I'm a specialist community physiotherapist. This summer I also qualified in specialist physiotherapy for children and adolescents.

In the spring of 2014, I completed postgraduate training in children's physiotherapy at Oslo and Akershus University College. As part of my degree, I wrote a case study, and that is what I want to present here. The article concerns spinal muscular atrophy and training in Innowalk as a supplement to traditional physiotherapy.

In this article, I shall present a case in which I have chosen to look at the effect of training in Innowalk for a boy with the muscle disease spinal muscular atrophy (SMA). I have compared the results with results from other studies that focus on training/physical activity for the group of neuromuscular diseases in general and SMA in particular (Carter, 2012, Grimby, 2004, Abresh, 2009, El van der Kooi, 2013).

Presentation of the case

Martin is seven years old and has a diagnosis of spinal muscular atrophy, type 3. He has reduced muscle tone, strength and balance, affecting his walking function, which is unsteady and rolling. He easily loses his balance and falls over, and it is difficult and requires a lot of energy for him to get himself up. Martin often has to sit down, and needs help to get up and to get dressed.

Over the last two years, his carers and other people around him have noticed a decline in his functional level. Martin confirms that he quickly tires when playing and doing activities at school and at home. The diagnosis is progressive in nature, and will in time lead to further loss

FACT BOX

Spinal muscular atrophy (SMA) is a hereditary neurological disease, the most important symptom of which is weakness in striated muscle tissue. The disease is one of the most common muscle diseases (neuromuscular diseases). Onset may be during gestation, in early childhood or in the teenage years, and there are significantly different degrees of severity (Frambu 2013).

SMA is defined in three groups:

Type 1 can only lie, starts before the age of 6 months
Type 2 can sit, starts before the age of 18 months
Type 3 can walk, or has been able to

of function (Campbell, 2012).

His physiotherapy treatment focuses on measures to establish good routines for contracture prophylaxis, increase stamina and independence, and maintain walking function for as long as possible.

The Norwegian Labour and Welfare Administration (NAV) provides assistance to obtain technical aids. Martin is still able to walk, but two summers ago I procured a manual wheelchair for him. He uses this to go longer distances and to get around quickly.
Prior to Innowalk, Martin was having weekly physiotherapy, trained three times a week with an assistant, and did daily mobility and stretching exercises with his carers.
In autumn 2013, I applied for a trial with Innowalk for Martin. The aim was to improve the effect of daily movement and stretching in the home, and to provide an alternative/supplement to traditional stretches. At times, it was difficult for Martin's parents to motivate him to do his training because he was tired. The hassle of following up training/physiotherapy every single afternoon was also an extra strain for both Martin and his parents.
Innowalk was intended to motivate Martin and make it easier for his parents to follow up on his training at home. A further aim was to prevent contractures, allow weight bearing on the joints in a 'more correct' position, maintain muscle strength and, as a result, enable Martin to retain walking function for as long as possible. Another goal was to provide Martin with motivation and joy of movement through physical activity on his own terms.

Theory

Spinal muscular atrophy (SMA) is a hereditary neurological disease, the most important symptom of which is weakness in striated muscle tissue. The disease is one of the most common muscle diseases (neuromuscular diseases). It may start during gestation, in early childhood or in the teenage years, and may have significantly different degrees of severity (Frambu, 2013). SMA is defined in three groups:

Type 1: can only lie, starts before the age of 6 months
Type 2: can sit, starts before the age of 18 months
Type 3: can walk, or has been able to

This classification provides a practical way to group the variations in the disease (Frambu, 2013, UNN, 2013). In type 3, the disability usually manifests before the age of ten, and includes problems getting up from the floor, using stairs and keeping up when playing with peers (Campbell, 2012). This is the case for Martin. According to Campbell (2012), the pathological characteristic of SMA is an abnormality in the large anterior horn cells in the spinal cord. According to Oslo University Hospital (2013), signals from the brain are not transferred to the muscles. The muscles do not function, leading to muscle wastage/atrophy. The number of cells lost and the progressive degeneration of the remaining cells correspond to the loss of function (Campbell, 2012).

From a perspective of socio-cultural participation, social participation is a prerequisite for human development, wellbeing and selfhood (Rogoff, 2003). Through social participation in culturally constituted communities, Martin will shape his personal development throughout his childhood and on into his adult life (Gulbrandsen, 2006).
Training is defined here as purposeful, systematic and regular activity to maintain and, if possible, increase muscle strength, physical function and stamina in hereditary chronic, progressive muscle diseases (ffm.no, 2013). According to UNN (2010), enjoyment of physical activity leads to a more positive attitude to one's own body.

Procedure

Before starting the trial with Innowalk, I gathered information from others with experience of SMA and Innowalk, and discussed my intentions regarding the user with several colleagues. In order to acquire this aid for Martin to try out, I had to carry out detailed preliminary work and obtain approval for the trial from both the Consultant Physiotherapist at the Rehabilitation Centre and the HSE representative. Before, during and after the trial, I recorded videos of Martin walking with and without the walking frame, standing up from a sitting position on the floor, and how long this took. The aim was to document any results/changes and to provide a basis for comparison (baseline).

Martin stood in Innowalk for at least 45 minutes, five days a week for four weeks.
An important element of the trial was logging the training every day, and I made a home visit once a week for follow-up. This ensured good compliance, in the sense that it made sure Martin was using Innowalk as intended, and made us all aware of how he was using it and the effect of the training. There were two days when he did not use Innowalk because he was ill

Results

Some positive results were seen after just one week. Martin said it was easier to go into his parents at night, and his mother had noted both that the training in the afternoons was going more easily and that Martin had lost weight. Half-way through the trial period, I observed that Martin's quality of movement when standing up/getting up from the floor had improved. Part of the reason for this was that he was now choosing to get up via a half-kneeling position. The video recorded after the trial period showed that Martin no longer needed breaks to walk the same distance without aids. The assistant said he had more stamina during the school day. His parents summarised the results as follows:

Martin doesn't complain about pains in his legs. We find he has improved circulation: his legs are nice and warm. He is crawling upstairs again, and has more energy on a daily basis. He has lost weight. The parents also say they can see that it is generally easier for him to move. Martin himself says that he 'runs' like other children. Daily training is going well. Martin is motivated and happy in his new aid.

The intention with using the aid in the home has been realised, and many of the other goals also

seem to have been achieved. It is interesting that the results were apparent after only a short time. Martin's muscles seem to be working better. His parents are both positive and optimistic about what Martin is getting out of the training; it does him good in several ways. Martin is happy in Innowalk and says that it is easier to move around. This is probably why the comprehensive training programme is going well.

Discussion:

Various studies of physical activity in persons with neuromuscular diseases (Grimby, 2004; Abresh, 2009; El van der Kooi, 2013; Wahl, 2009) support several of my findings. Physical activity and training can counter inactivity and secondary effects such as overweight, osteoporosis (brittle bones) and cardiovascular disease, as well as contributing to increased energy (Grimby, 2004). In Martin's case, we saw weight reduction after just one week, and he quickly gained more energy.

According to Campbell (2012), traditional training principles explicitly state that achieving an improvement in fitness requires one to train a minimum of three times a week and maintain this for eight weeks or more to see results. Several small studies have reported improvements in walking, strength and function following treadmill training for children with CP. I assume parallels can be drawn with Innowalk, and therefore believe that improved walking function and increased energy in daily life may be a result of improvement in fitness. The training principles specified above are satisfied (45 mins x 5 days per week), although the duration has not yet passed eight weeks. Martin will continue to be followed up on a weekly basis at school and regularly with regard to Innowalk. I believe this will reduce the risk of over-training, which brings us to the next point.

In the case of diseases of the motor anterior horn cell – here SMA – there is a concern that too much training can increase the loss of the muscle-controlling nerve cells (motor neurons). The healthy nerve cells can become overloaded if they try to compensate for the lost cells (Abresh, 2009). It is important that Martin is not exposed to over-training, which in turn can bring about a form of fatigue syndrome. The training 'dosage' must therefore be taken into consideration. Training without pain is the key, according to paediatric endocrinologist Alfred Slonim (Abresh, 2009). His blueprint for training is short and sweet:

'No gain with pain'. He recommends light aerobic training such as sessions on the treadmill and cycling, all supervised by a doctor/physiotherapist with the requisite knowledge (Abresh, 2009). Both treadmills and Innowalk are motorised forms of training equipment, which is why I consider training in Innowalk to be ideal if Martin does not experience pain at the time or afterwards. He has not indicated any pain while training. Pains immediately after or a day or two after training are a sign of incorrect training or over-training, and should result in the training being stopped temporarily or reduced in intensity (Abresh, 2009). Martin's parents were informed of this so that they could provide quick feedback if pain occurred in connection with or subsequent to training.

Martin no longer has pains in his legs, and I believe the regular movement he has experienced using Innowalk at home may be the reason for this. Abresh (2009) writes that correct training may reduce some forms of pain. If the pains are an effect of stiff joints and long-term immobility, training in the form of stretching and joint mobility may be useful (Abresh, 2009).

I believe that maintaining independent walking function for as long as possible will delay the development of contractures in the joints, as well as the development of scoliosis. This is confirmed by Campbell (2012), who writes that individuals who maintain independent walking function have a lower incidence of scoliosis and less serious curvatures if scoliosis should develop.

It is important to emphasise Martin's own experiences in connection with the training. After a short time, he said – in his own words – 'I can run like a normal child' and 'training in Innowalk is good'. It is important to engage Martin so as to maintain his motivation to continue training, and to provide affirmation.

All action and interaction is participation. I consider that maintaining Martin's walking function for as long as possible may help to ensure his social participation at school and in daily life. It will be easier for him to follow his classmates around. By taking part in organised experiences of everyday life and in opinion-forming processes over time, Martin will develop as an individual (Gulbrandsen, 2006). I believe that continued walking function will strengthen his potential for participation: participation with regard to his disability, in an individual and social perspective (Grue, 1999; Ulvik, 2009).

If the results continue in this positive direction, I think this may be very good for Martin. An improved functional level will give him a sense of mastery, which will help to improve his self-image and self-confidence (Grue, 2013).

I consider my experiences in this case may be significant for others with the same diagnosis as Martin. Innowalk enables any walking function and general physical fitness to be maintained and possibly even improved. This, in turn, will have positive ripple effects, as described above.

Conclusion:

The case report has shown that for Martin, who has SMA, training in Innowalk has produced positive results in a short time in the form of better walking function, more energy and a genuine sense of moving more easily. It remains to be seen whether the results are lasting. Eighteen months after starting to use Innowalk, Martin is still very happy in it and continues to have better sitting balance, trunk control, arm strength, etc.

The time perspective of the case was limited, but I chose to be positive since several of the results are supported by various studies of training in people with neuromuscular diseases (Grimby, 2004;

Abresh, 2009; El van der Kooi, 2013; Wahl, 2009). If the training is adapted to the individual child, takes account of his/her particular needs and requirements, and is closely monitored to avoid over-training, I think that Innowalk can be recommended for this user group. Grimby (2004) found that strength and conditioning can be carried out with a positive effect. The condition for this is that the training programme must be drawn up individually, based on analysis and knowledge of the diagnosis and functional level, and adapted on the basis of an understanding of the underlying pathology. Grimby (2004) states that the effect of the training must be monitored carefully.

Several health care professionals are seeing the benefit of Innowalk in their practice. It would be interesting and useful if more colleagues were to document their results and confirm any effects of using this aid. Systematising the information in this case report has made me aware of what, how and why I have thought as I do, and given me new ideas for further work with Martin and other children I meet in my practice.

The goal of increasing Martin's motivation for training at home, and improving how this goes, has been achieved – despite the fact that training in Innowalk is a time-consuming activity. However, I believe Martin and his parents will be motivated by seeing the results of the work they are doing, in the form of an improved functional level for Martin.

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