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Dynamic Postural Management for

Progressive Neuromuscular Disorders in Children and Young People and Case Studies

A Clinical Application Paper by Aergo Health

2022 Edition 1

Contents

4

Progressive Neuromuscular Disorders and Postural needs

Neuromuscular Disorders (NMD) can be defined as "a range of conditions that impair the functioning of the muscles, either directly, being pathologies of the voluntary muscle, or indirectly, being pathologies of the peripheral nervous system or neuromuscular junctions" (RCN, 2022).

Some NMDs are progressive, meaning the symptoms worsen over time. The most common progressive NMDs are Muscular Dystrophy and Spinal Muscular Atrophy, which are both genetic disorders that cause muscle weakening over time.

Muscular Dystrophy (MD)

MD is an umbrella name for a group of muscle diseases caused by gene mutations. Over time, muscle weakness decreases mobility and function, making everyday tasks difficult. In later stages, there may be difficulties with respiration, digestion, and heart functions. The muscles affected, and the severity of the symptoms vary with the type of MD. Some types of MD are more prevalent in males, and some affect males and females equally. The age of onset varies from babies to adulthood (MDUK, 2022).

Duchenne Muscular Dystrophy (DMD)

This is the most common type of MD which occurs approximately once in every 3500 male births. Duchenne symptoms may be detected from two years old, starting with weakening of the lower limbs. Children with DMD begin to develop a waddling gait and struggle with balance, often losing independent mobility between 8 to 12 years (Sienko Thomas, Susan et al., 2010). Children with DMD have a reduced life expectancy.

Spinal Muscular Atrophy (SMA)

SMA is a genetic disease caused by the loss of motor neurons in the spinal cord. This nerve cell loss affects the nervous system and the control of voluntary muscles. The most common form of SMA is chromosome 5 SMA which varies in age of onset from birth to adulthood. The earlier the onset, the greater the severity of symptoms and rate of progression. Children with symptoms at birth or in infancy (Type 1) tend to be most severely affected. Adults (Type 4) have the highest levels of motor function (MDS, 2022).

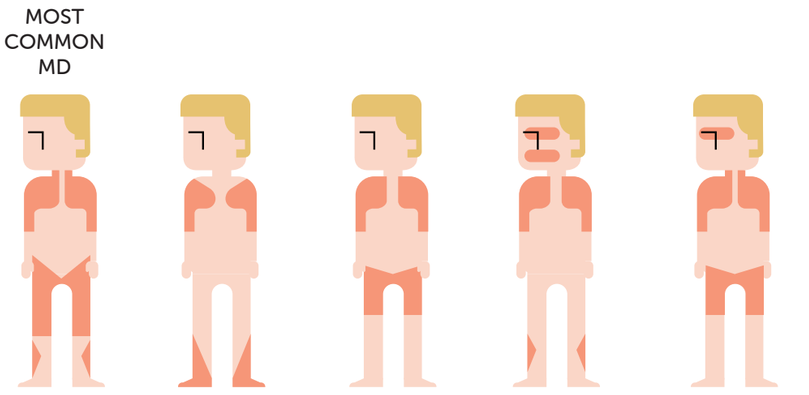
SMA Types 2 and 3

An onset of symptoms between 3-15 months is known as SMA Type 2. Children usually learn to sit independently but lose this ability later. They are unable to stand or move without support as weakness begins before they gain these skills. Onset between 18 months and 18 years is known as SMA Type 3. Children generally achieve independent mobility but may fall frequently and have difficulty climbing stairs. They may later lose the ability to stand or move independently.

Affected muscle areas

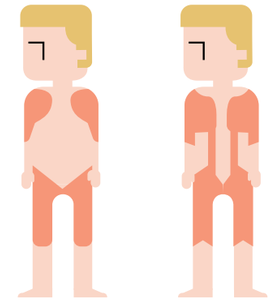
Highlighted in red

Muscular Dystrophy



MDs are caused by gene mutations which impact the production of essential muscle proteins, causing muscle weakness. MDs are diagnosed by identifying the location of the affected.

Spinal Muscular Atrophy



SMA is caused by a deficiency of Survival Motor Neuron (SMN) protein, and effects weaknesses in muscles closest to the centre of the body: shoulders, hips, thighs, and upper back.

SMA Type 0, Type 1, Type 2 (intermediate SMA/Dubowitz disease), Type 3 (Kugelberg-Welander disease), Type 4

Specialist Equipment

Children with MD or SMA require a range of specialist equipment as their condition progresses. This includes a wheelchair for mobility (Thais et al., 2017); special seating to provide postural support for the pelvis, trunk and head; and a standing frame to counteract the effects of prolonged sitting. Support in this equipment can improve upper limb, respiratory and digestive functions(Archer et al., 2016). The management of these conditions is complex and relies on the input of a large multidisciplinary team, of which specialist equipment forms an essential part.

For further reading on Neuromuscular disorders, you can find a list of suggested clinical resources on page 16.

6

Dynamic Postural Management With Aergo PS

Specialist seating is used by children living Muscular Dystrophy (MD) and Spinal Muscular Atrophy (SMA) as an essential tool for providing comfort, maximising function, and helping to maintain postural alignment.

Static Seating

Most of the adaptive seating available for children with NMD provides static pelvic and trunk support, meaning that little active movement is possible within the seating system. Direct feedback from parents of children with MD and SMA indicates that due to low muscle tone, a child can move out of the prescribed position and struggle to return to the original position because of muscle weakness. Harnesses and straps may be introduced to secure a child's posture, but this often does not resolve the problem, instead becoming restrictive and causing discomfort over time. In addition, modular seating systems achieve their intended aims best when the user matches and remains correctly in the prescribed setup.

Humans are active by nature and movement is essential for exploring and understanding the world around us. Movement also helps to train functional movements and provide sensory input (L. Lange et al., 2001), which is critical to the development, independence and quality of life of the child.

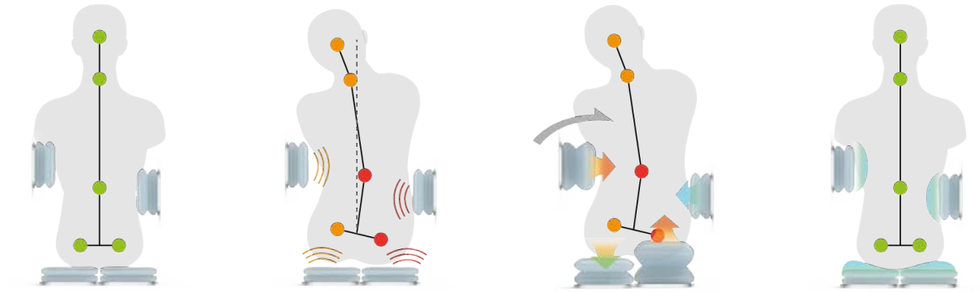
Opportunity for New Innovation in Seating and Postural Management

There is a wide variety of robust and clinically validated seating systems available on the market, yet the majority offers static support. The user is unable to change positions actively and is highly dependent on posture facilitated by the seat (Strobl, 2013) as well as the correct placing of the person into the seat by carers. The user's physical positioning becomes highly dependent on the clinician's initial prescription, and carers' understanding of why the position is important.

In our changing healthcare world, with clinical and wheelchair services still trying to catch up on a backlog caused by the Covid-19 pandemic, access to services provided by these specialists is less readily available. Families report that appointments for assessment involve lengthier waiting times, and reviews for fitting, adjustment or replacement take longer than anyone desires.

To address the need for children to receive timely equipment provision, and the desire to provide less time-intensive follow-up and monitoring, Aergo Health set out to answer the following questions:

- **Can we create a seating system that continuously moves with the child while maintaining a healthy seated posture?**
- **Can we facilitate remote postural management?**



Starting point: Prescribed setting by clinician ▶ Sensing postural deviation ▶ Adjust inflation to correct deviation ▶ Monitor and reactive management

Aergo Health's answer: Dynamic postural management using pressure sensitive air modules

In response to both the needs of children and clinicians, Aergo Health has created and patented a dynamic seating solution using pressure sensitive air cells. These highly responsive air cells continuously monitor how a child is seated and automatically adjust support levels to promote the optimum seated position identified during assessment.

The technology works by recording the child's starting position, determined by an occupational therapist or physiotherapist during the initial seating assessment. The setup of the air cells is saved and safely stored as a fixed setting in the Aergo PS control app.

As the user changes position, the air cells sense pressure changes and use this data to determine the extent of change of the child's posture. Dependent on the user's positional changes, each air cell adjusts support levels either by inflating or deflating to gently prompt the user back to their prescribed functional position.

The air cell system continues this cycle of analysis and adjustment to manage the user's posture without restricting the user's movement. The user or the carer also can make minor adjustments when a clinician is not available to ensure the user is comfortable.

The Aergo PS control app provides a simple control interface which may be used independently by users who are assessed as competent to make simple adjustments to the air support levels. They can also set up virtual calls with their therapists, allowing specialists to conduct remote postural review and baseline adjustment.

8

Case Study:

Dynamic postural management for young wheelchair users living with progressive neuromuscular disorders

This case study was written following the structure recommended by the Royal College of Occupational Therapy UK.

Background

Children with Muscular Dystrophy develop muscle weakness which limits their ability to sit in an upright posture. They rely heavily on specialist seating systems and interventions from carers and clinicians for repositioning because independent postural control is fatiguing and difficult to sustain. Without a seating system to provide external support, gravity exacerbates spinal deformities which impacts on respiration, digestion, and independence.

Specialist seating systems are widely prescribed by therapists to support children with MD in an optimal seated position. These solutions reinforce a stable and balanced position of the pelvis, trunk and head, facilitating upper extremity function. This is critical to a child's independence in completing simple daily activities (Green and Nelham, 1991). Although specialist seating systems are effective in maintaining postural control, their static nature can become restrictive to a child's movements (Green and Nelham, 1991). Children are active by nature and often move out of the prescribed seated position. Best practice in postural management is known to include rest, recreation, and the active positions a child may adopt throughout the day. Thus, prolonged static posture can impact negatively on a child's physical function (Kinali M et al.,2007).

A dynamic seating system which reacts to shifts in position is needed to ensure optimal posture is maintained without restricting active movement and compromising independence.

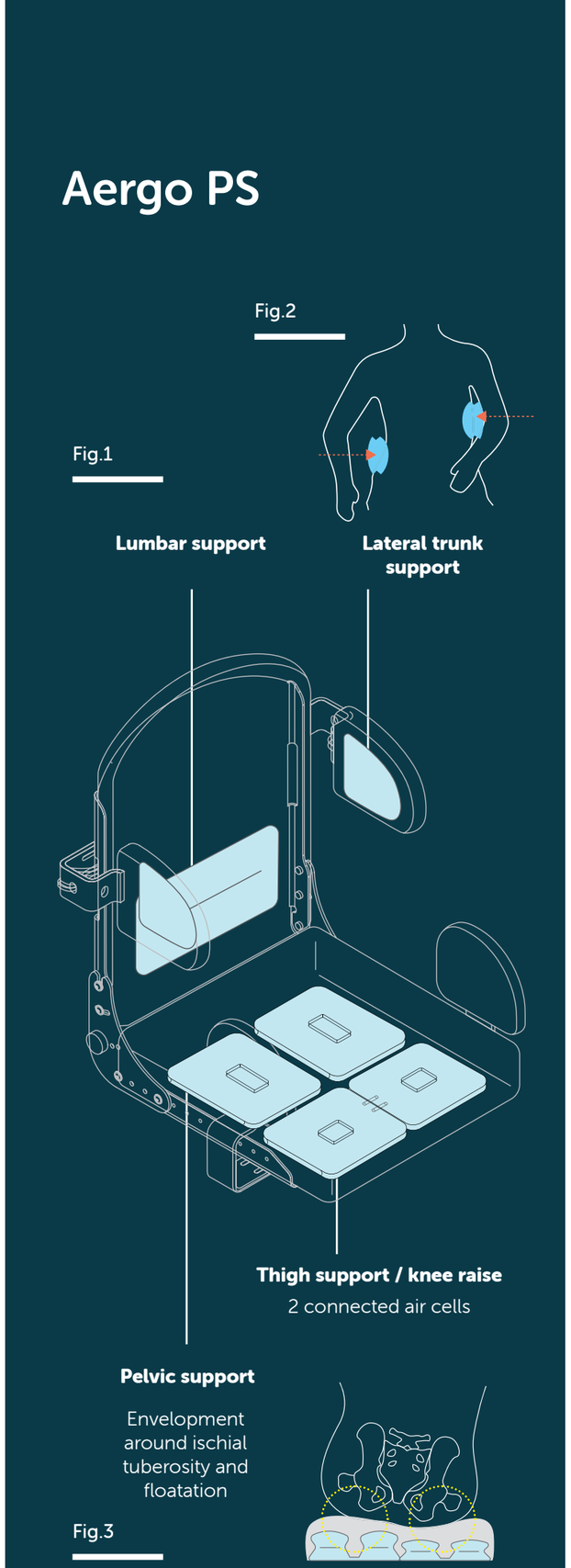
Intervention

Aergo Health has developed a dynamic seating solution, the Aergo PS, to accommodate the changing postural needs of children aged 6-16 years old with neuromuscular diseases.

The Aergo PS is designed as a dynamic seating system that replaces foams and wedges to support a child's constantly changing posture.

Aergo PS uses pressure sensitive air cells that are reactive to movements. Each air cell offers 5 levels of inflation, where 1 is the lowest and 5 is the highest. Without any user weight applied, the air cell has an adjustment range of 0 to 10 centimetres. The accompanying control app (available on android and iOS platforms) enables clinicians to provide hands off positioning during postural assessment and prescribe the optimum support settings quickly. The user can then independently fine-tune support levels for postural support as their needs change during prolonged sitting, but safely stay within the clinician's predetermined parameters.

Once the system is set up according to the clinician's assessment, the Aergo PS can react to changes in position based on pressure data deviations and automatically adjust individual air cells to gently assist the user back to the prescribed optimum seated position.

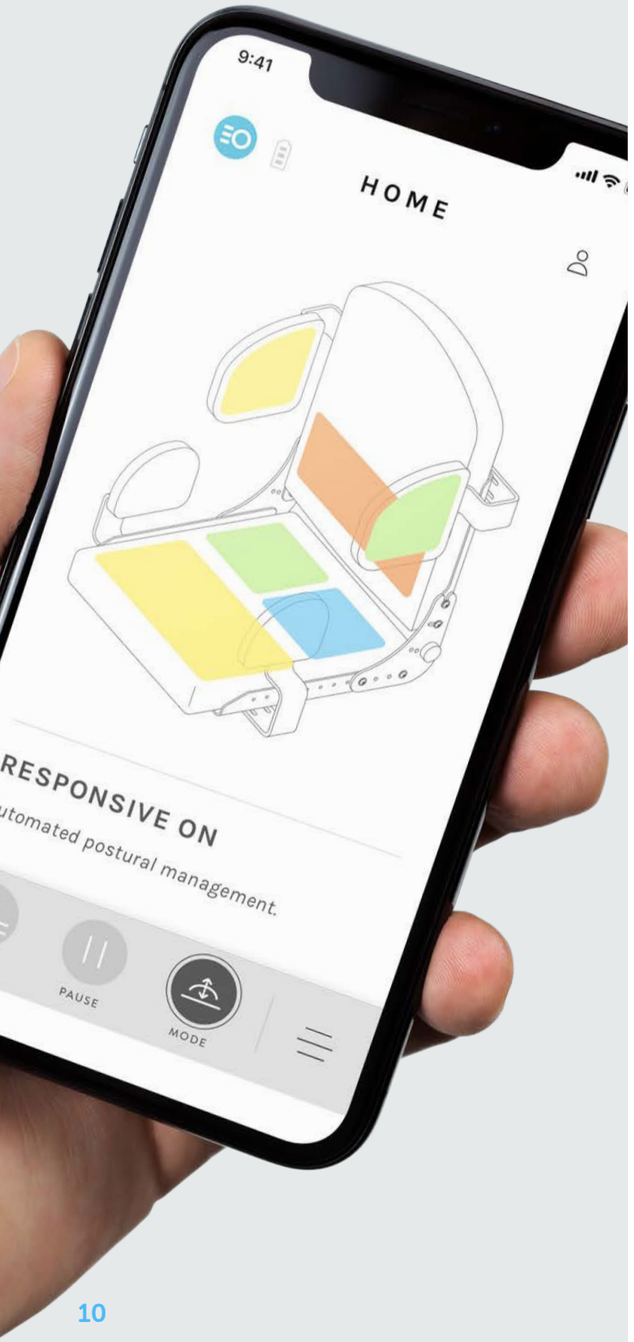


The system comprises of six air cells (Fig.1) positioned across the seating surface, offering upper trunk, lumbar, pelvis and thigh support. Each air cell is independently adjusted depending on the user's seating needs. The shape of each air cell was designed using the anthropometric data of children and young people aged 6-18 years. When inflated, the lateral and lumbar air cells contour to the user's thoracic (Fig.2) and lumbar curves. Pelvic air cells offer both envelopment and flotation of the ischial tuberosities (Fig.3), key support measures to provide stability and diffuse pressure. The thigh support air cells offer immersion along the femur. Height adjustment under the femur helps to maintain pelvic stability and reduce shear forces which occur through sliding forward in the seat. In combination, the system can be purposely tailored to accommodate a wide range of posture quickly.

As users fatigue and lose posture, the air cells automatically adjust inflation levels to reposition them.

The design and postural management principles of the Aergo PS are based on recommendations from specialist occupational therapists at Alder Hey Children's Hospital and established publications such as Seating and Wheeled Mobility and MDUK Wheelchair Guideline. A pilot study was conducted to test the design and postural management principles in the real-world, so Aergo Health can effectively support the seating needs of children with MD.

Aergo PS App



Outcomes: Evidence of Impact

A 3-month home-based pilot study was conducted with two children with Duchenne Muscular Dystrophy (DMD). Set up of the Aergo PS was discussed with the participants, their parents, and occupational therapists.

Child A Initial Set-up

Child A's Aergo PS was installed on a manual wheelchair. The Aergo PS was set up based on the initial postural assessment, and evaluation of the child's environment and their activities. Child A has weak neck and trunk control and naturally sits in a kyphotic posture. He has experienced occasional lower back pain due to lack of lumbar support on the backrest. The occupational therapist started by ensuring the frame of the Aergo PS was set up correctly for Child A's symmetry, then Child A began independently adjusting the air cells following instructions from Aergo Health. He started with inflating both lateral air cells to level 2 which provided sufficient support to keep his upper trunk in the desired position.

Child A described the feeling as being "hugged into place".

He then inflated the lumbar support and pelvic supports to level 2. After 2 minutes, Child A could feel a significant push from the air cells while the occupational therapist could see that Child A was being pushed forward in his wheelchair. The occupational therapist repositioned the lumbar air cell to the lower section of the backrest facing the upper pelvic region and deflated to level 1. This setting was more comfortable for Child A and it was sufficient to generate a small amount of posterior pelvic tilt when observed from the sagittal plane.

Child A inflated both pelvic air cells and thigh support to level 2, the occupational therapist reached under the side of his pelvis to feel the support and inflated the left air cell to level 3 to accommodate his pelvic obliquity. Child A was comfortable with the set up and his posture appeared neutral. The setting was saved to the mobile app as the pre-set default.

For child B, the Aergo PS was set up as a floor sitter. The PS frame was set up to match the child's body measurement and the air cells were inflated using a collaborative process between the occupational therapist and the child. Child B has a weak trunk but can support himself during the day in sitting. He has knee contractures and uses a walking frame to move around the house. He also uses leg gaiters in the evening to provide stretch. Child B is in the process of obtaining a new wheelchair as his muscle strength has deteriorated. Aergo PS was set up on the floor of Child B's living room, where he spends most of his evening watching TV and playing XBOX with his sibling. Child B's occupational therapist inflated the right pelvic air cell to level 2 and left pelvic air cell to level 1 to accommodate his pelvic obliquity. Child B's right lateral was inflated to level 3 and left lateral to level 1 to gently correct his scoliosis. Thigh support was inflated to level 2 which created more stability on the seat. Child B now sits neutrally on the PS and is comfortable with the setting, thus a pre-set default setting was created.

Monitoring

Every two weeks, visits to both children were conducted to collect feedback from them and their parents. A Likert scale questionnaire was used with each child to collect quantifiable data about their experiences using the Aergo PS. This questionnaire is validated for use with children (The Five Degrees of Happiness, TFDH 2), and asks participants to rate their enjoyment, independence, posture and comfort while using the Aergo PS. In addition, observations and qualitative interviews were conducted to further capture each child's experience and feedback.

Feedback from the Child A

Child A reported that he deflated the air cells in the seat by 1 level as he felt it was raising his feet from his foot plate. He also reported that he deflated the lateral air cells when he put on a thick sweater at home. He found the ability to make the adjustment very convenient. In the evenings, the PS was moved to Child A's bedroom and used as a floor sitter while Child A played computer games. He enjoyed the set up as he felt the PS gave him a lot of support while he held his controller. He also found it comfortable while wearing leg gaiters in the evening.

Child A had not triggered the cycle mode for comfort, as he found the default dynamic movements of the air cells and the softness of the cushions sufficiently comfortable during prolonged seating.

Feedback from the Child B

Child B previously sat on a bean bag that did not provide any support, and he would sit for over 2 hours in front of the TV in the evenings slumped in the bean bag. With the PS, Child B sat comfortably in a neutral position for the entire duration of his home-based activities – an estimated 4-5 hours. Child B reported liking the PS most as a floor sitter and enjoyed the thigh support which prevented him from sliding forward. Child B triggered the cycle mode weekly as he enjoyed the gentle movements across the seat. Notably, Child B's parent remarked that he played more online games during the Aergo PS trial.

Parent feedback

Parent feedback consistently highlighted the reduced requirement for physical repositioning of their child's posture. After 3 weeks of the children using the device, both parents felt off-loaded and confident with the device supporting their children's postural needs without supervision.

When asked if the Aergo PS had impacted on the child's postural management, Child A's parent stated that "when sitting in the PS he is more mindful of his seating position. He will adjust cells in Aergo PS to assist better seating".

Child B's parent commented that he sat in the Aergo PS every day when he did his homework or played computer games. His parent stated that "Yes, he is so much more comfortable and supported than before. Him sitting for at least 2 hours a day and keeping a good posture is doing him good. Without the PS he starts to slouch but with the PS he can adjust himself. He never complained." His parents felt that because the PS gave Child B better support, he was more engaged with online gaming which enabled him to make more friends. This was important because Child B struggled with social activities in school and felt isolated due to his disability.

> "He enjoys it because of the feeling of ownership, control and able to change different levels of his posture without relying on me or others."

At the end of month 3, both users rated the device as highly enjoyable to use (5/5), supportive of their postural needs (4.25/5), having improved independence (4/5) and comfort during prolonged seating (4.25/5). Based on their interviews, the children stated the reason for enjoyment and comfort was the ability to make small adjustments to the support level instead of feeling fixed in a single position which often led to discomfort.

14

Digital Health Trends In Occupational Therapy & Aergo Health's Future Development

Presented as a poster presentation at the Posture and Mobility Group (PMG) Conference 2022

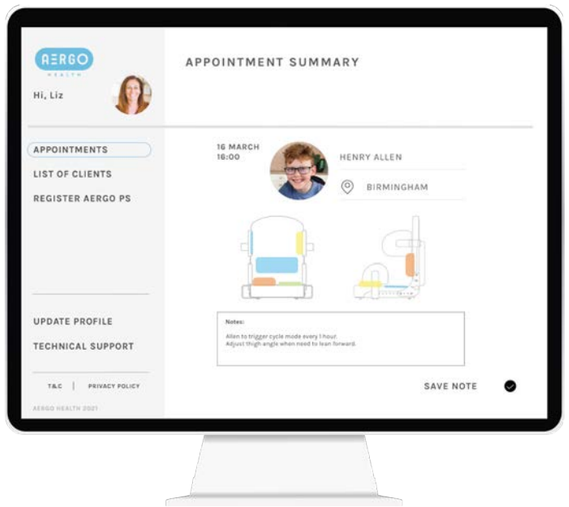
The rise of Telehealth and future potential for occupational therapy

The impact of COVID-19 has led to a decrease in healthcare access for existing and new wheelchair users who are categorised as at high risk of developing severe illnesses from the virus. In the UK, staff shortages across the NHS have further exacerbated accessibility to clinical support. In order to overcome this, conference calls and telehealth technology have been widely adopted to provide care and treatments remotely.

An international survey with over 1000 therapists has found the pandemic has increased the integration of telehealth in their clinical practices by 66% (Chantal Camden & Mindy Silva 2021). Video platforms such as Zoom and Microsoft Meet are commonly used for remote patient-screening and follow-up appointments. Although further studies have shown that the increase of telehealth usage has improved patient satisfaction and caseload managements, not all physical assessments are easily replicated remotely.

Wheelchair and seating prescription are intrinsically complex and require therapists to conduct extensive in-person assessment in order to provide the correct equipment. Often, finding the right postural support for paediatric wheelchair users is especially challenging as their bodies and needs change so quickly. Findings from an equivalency study in Telerehabilitation for wheeled mobility and seating assessments found that effectiveness between in-person and telehealth treatments were highly comparable (Schein et al., 2010). Leading wheelchair manufacturer, Permobil, stated that 10,000 users of their power wheelchairs have activated internet connection in their wheelbases for remote technical diagnosis. Research and Development in internet connected specialist equipment is on the rise, and the benefits of connecting clinicians and trained technicians to users' wheelchairs and equipment will benefit remote clinical support significantly.

Fig. 3 Aergo Clinician Portal Appointment Summary



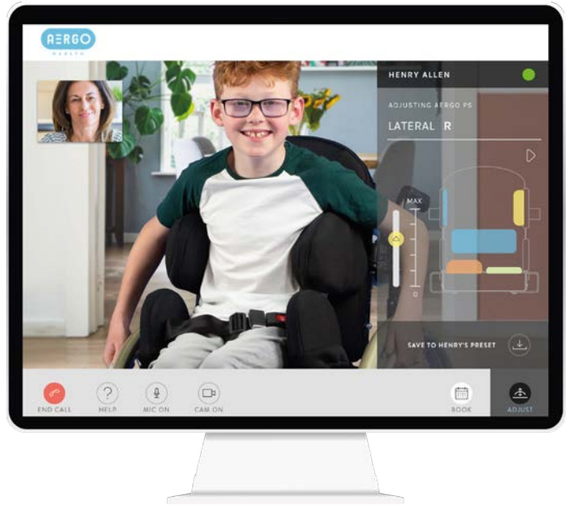
Digitising postural assessment

The pandemic has pushed Aergo Health to rethink how we can better service our clients and prepare for the change across healthcare provision. For years we've heard clinicians talk about the difficulty with case load managements and the shortage in staffing. We've also heard clients talk about not being able to see their clinician for extended periods of time. A combination of these issues and the change in perception in digital health poses an opportunity for change. Aergo Health decided to develop a digital postural management portal for therapists to remotely assess client's seating needs and make real-time adjustment to client's posture. Our objective is to enable therapists to have a high level of intervention through a digital control interface coupled with internet connected postural support air modules.

Identifying user needs

Primary interviews were conducted with wheelchair service occupational therapists through conference calls. A series of open questions were asked to understand the changes in the therapist's clinical practice and experience with integrating telehealth into assessments and routine check-ups with clients. The qualitative feedback provided insights that led us to create user requirement documentation and informed our software feature list.

Fig. 4 Aergo Clinician Portal Conference Call



To bridge the gap between digital and in-person assessment requirements for wheelchair equipment provision, Aergo Health has developed a clinician web portal that connects therapists to their client's Aergo PS seating system. The web portal (Fig.4) is designed to facilitate collaborative postural management between wheelchair users, carers and their therapist. The Aergo Clinician web portal provides a unique conference call facility that not only uses video call features to capture a user's seating position and feedback, but also visually displays the user's real-time pressure data to inform sitting position and comfort of the user. A user-friendly control panel is included to enable real-time adjustments of individual air supports within the client's Aergo PS.

Result & Testing

Observational studies have been conducted between therapists and wheelchair users to evaluate usability and stability of the Aergo

Clinician web portal. The result demonstrated ease of use and effectiveness in fine-tuning a user's postural support system to ensure the equipment set up continues to support the client effectively.

The Discussion

Although observational studies have demonstrated promising potential of digital postural management by enabling remote control of the Aergo PS system through the web portal, there are still limitations particularly with initial equipment set-up and body measurements for clients with complex seating needs. Initial feedback has suggested the key benefit of the web portal in clinical practices is the ability to track adherence and make minor adjustments to the equipment quickly and frequently, whilst reducing the need to travel for a physical meeting. Aergo Heath will expand the data points for therapists to accurately conduct remote assessment and rehabilitation. Future development could include diagnostic tools using Augmented Reality(AR) body mapping and integrated high fidelity pressure mapping to better inform the user's sitting position.

The future development of technology at Aergo Health aims to create a digital postural management assistant for therapists to provide effective digital postural and pressure care for their clients.

18

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Further reading

Wheelchair Provision for Children and Adults with Muscular Dystrophy and other Neuromuscular Conditions

Access at https://www.wheelchairmanagers.org.uk/downloads/Wheelchair%20Guidelines.pdf

Seating and Wheeled Mobility: A Clinical Resource Guide

Available for purchase at https://www.slackbooks.com/seating-and-wheeled-mobility-a-clinical-resource-guide/

A Clinical Application Guide to Standardized Wheelchair Seating Measures of the Body and Seating Support Surfaces

Access at http://www.besrehab.net/news-articles/a-clinical-application-guide-to-standardised-wheelchair-seating/

"Let's Get It Clear" series by Dr. Barend Ter Haar

Let's get it clear: Early Intervention – the needs of younger people
Let's Get It Clear: Dynamic Seating – What does it involve?

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