Keywords: Assistive Technology, Dystonia, Collaboration.

Abstract
This is a case study of the occupational therapy rehabilitation process of a teenage girl who presented in 1997 with a rare neurological condition diagnosed as rapid onset Dystonia Parkinsonism. She was helped to obtain some of the occupational performances of her choice by assistive technology devices tailored to her needs by the rehabilitation engineering department, and in this paper we wish to point out how collaboration between our departments can be of benefit to patients, and how essential microelectronic technology is in the occupational therapy environment.

Referral
A thirteen year old was referred in March 1998 to the rehabilitation hospital. She had been transferred from a children’s hospital where she had spent 5 months, having presented with acute onset of dystonic spasms throughout her body musculature.

Social Situation
She had been, up to the date of onset of illness, a normal, healthy 13 year old, commencing her junior certificate programme.

Medical History
A previously healthy 13 year old girl with a family history of neurological illness (dystonia), developed pain in the fifth digit of her left hand. This progressed to pain and stiffness in the arm. Within a week she was suffering frequent pain and spasm in the left arm which she held flexed, and by this time she was having difficulty with speaking and was drooling. Her legs also developed stiffness and this led rapidly to inability to walk. Tongue, lip movements and swallowing difficulties also occurred. CT and MRI of brain were normal. Vision, sensation and cognition were normal, and she remained continent of bowel and bladder. Medication included valium (Diazepam) and Lioresal (Baclofen) and phenytoin.

Occupational Therapy.

Observations on initial assessment

Physical Aspects
Referral to occupational therapy occurred pre-admission and her first attendance was as a day patient. At that time she was extremely fatigued and suffered from pain in the back and shoulders and in her right hand which had also started to show signs of increased tone – the elbow was flexed and there were intermittent painful spasms in the flexed right hand; tonal increases were exacerbated by effort but she had some ability to write tiny letters and to point to a tiny alphabet chart for communication. The left arm was in a pattern of total flexion, with no purposeful movement. The palmar skin and nails were affected by this and she had had problems with infection. Tongue projection, drooling and inability to close the lips were a feature also.

Self Care
She was dependent for all her personal care needs – dressing, toileting and bathing, and was on nasogastric feeds. Shortly after her day assessment she was admitted to the rehabilitation hospital.

Treatment planning
Occupational therapy assessment and intervention could be considered under the rehabilitation, biomechanical and neurodevelopmental frames of reference, the former being the prominent. Areas that required immediate intervention included:

- Corrective seating
- Splinting
- Pain management
- Functional performance in occupations important to her, i.e. communication, education and leisure as well as self-care.
- Identify her strengths, interests and resources through ongoing evaluations together with therapy.

It was important with this girl that assessment was continuous during treatment due to the changing nature of her condition and significantly the positive response she made to medications.

**Corrective seating:** Wheelchair adaptations were temporary and she received her own wheelchair with tray, which provided adequate support. She had good balance while sitting unsupported on a plinth but used the wheelchair for mobility and all functional performances. After approximately six months she progressed to walking with an Atlas walker, using a splint on the left arm and supervised continually. The wheelchair continues to be necessary in all other situations.

**Splinting:** was used for corrective purposes and for maintenance of maximum range; for pain relief and prevention of further decrease in range leading to skin breakdown and joint pain. Remouldable, thermoplastic, anti-spasticity splints were provided for the right elbow and hand and for the left elbow: she could only tolerate a sheepskin pad in the left hand.

During the rehabilitation period these have been reviewed and she recently has only needed the left elbow splint whilst walking (as above).

**Pain management:** the team felt that increases in the doses of valium played the most significant part in relieving painful dystonic spasms. This continued to be in association with Lioresal (the phenytoin was discontinued.)

At times of increases in tone - (sometimes this was in spite of resting or relaxation, at other times it was associated with increased effort.)- the girl used eye contact and direction of gaze to direct the therapist to open either hand gently and massage it -this would relieve the situation for some varying time. Otherwise she adopted her own strategy of biting her fingers which seemed to give her relief but was a concern to staff.

She spent some of her occupational therapy sessions in the multisensory room which made a considerable difference in the early stages, especially, incidence of painful spasms decreased and she commented herself that this was due to distraction by the various attractions in the room. It continues to be a favourite place for her to spend time in and the padded floor and walls allow her to experiment physically more freely and expressively, laughing and relaxing with obvious psychological benefits.

**Functional occupational performance and identification of strengths.** Assessments were done with various body parts to determine which were associated with less effort, tonic increases, and pain. At that time, very painful episodes of dystonic spasms occurred regardless of her efforts, but efforts also exacerbated this. On some days, fluctuation in fatigue and frequency of spasms could preclude any work with switches. She could control a switch with her knee, cheek, or right hand squeeze – which she preferred but was inclined to increase flexion spasms. She attained fairly consistent control of two switches placed at the lateral aspects of her knees, using active hip abduction.

The hospital’s rehabilitation engineering department was consulted and collaborated in assessments. Voice control – using four vocalisations – was tried, but after some rather fatiguing training sessions was discontinued as a consistent vocalisation could not be maintained. The engineer then made a joystick mouse alternative for control by her right foot which had emerged as her best method of control. This was moderately successful and she could now create art on her computer and loved this activity. At the same time she was practising painting with a brush taped to her right foot, and playing board games with her toes. However the engineer felt that an enlarged twelve switch mouse alternative would decrease the level of effort and made this for her in collaboration with the occupational therapy carpenter – this was fairly heavy and sturdy to take robust kicking but also responded to light touch by the toes or heel. She was thus enabled to make art and to type.

After approximately two months she started to regain more recovery in the right hand to the extent that she could use a keyboard in the vertical position with the index finger. Tone continues to decrease in the right hand and she can write better, uses pencils for art work and engages in craft work and baking activities using the right hand only.
She has received a Cameleon communication aid with voice output which is attached to the tray of her wheelchair. She walks with an Atlas walker with the left arm in a splint, and supervised. Balance remains unreliable and although she can feed herself once set-up, she is dependent for washing, grooming and dressing. Discharge plans include a move to a new house with a purpose-built extension and attendance at a school catering for physical disability where her present educational programme and her therapies can continue.

There follows a narrative account of the collaboration between occupational therapy and the rehabilitation engineering to exploit this girl’s changing abilities with assistive technology changing apace.

The rehabilitation engineering department provided support in terms of assistive technology to supplement and augment her functional abilities particularly with regards to education and communication.

**Letter-scanning**

Initially using the two-switch scheme operated using active hip abduction outlined in the identification of strengths section, an automated letter-scanning scheme was realised in software for use as a communication aid. This system (called the NatterBox) was simply a computerised version of the conventional letter board, the layout of which was designed by the speech and language therapist involved. The software program was written for the Windows 95/98/NT platforms and utilised the serial port for switch interfacing. Care was taken to minimise hardware so that the system could be ported easily between various computers (the two switch interface uses just three pins on the serial port, the single switch interface, just two). The software program itself and the wiring details can be downloaded from the National Rehabilitation Hospital (NRH) website. Figure 1 shows a screenshot taken from the program illustrating the letters organised in a column-row format along with the text box upon which the phrase is constructed.

![Figure 1](image_url)

The letter selection method varies depending on whether one or two switches are used. In single switch mode, operation begins with the screen of Figure 1 and the highlighting of each letter in turn in the first column for a time period tailored for the user (and which can be adjusted from a pull-down menu). Once the location of the required letter is known the user presses the switch when the row corresponding to the required letter is reached and scanning then proceeds on a row basis from left to right. Once the desired letter is highlighted pressing of the switch automatically places this letter into the text box and so, in this way phrases can be built up on a letter-by-letter basis. This scanning scheme, though initially adequate, did not fully exploit this girl's ability to interact with her surroundings as it soon became apparent that it was possible for her to control at least two switches. In order to exploit this extra degree of freedom a two-switch scheme was incorporated into the software so that row selection was done with one switch leaving column and letter selection to be controlled by the other. This scheme improved phrase construction time immensely. This reduced her frustration which resulted in less tone and so longer, less-fatiguing and more fruitful sessions could be achieved. More advanced versions of the software included the provision to assign often-used phrases to single buttons (so that they were available quickly when required) and speech synthesis capability.
Alternative Computer Mice

The girl after responding well to treatment progressed to the stage where she could avail of more advanced interfacing strategies; one such method involved the use of speech-recognition software as a command mode interface to a PC. This idea proved more difficult than anticipated as vocalisation repeatability was poor and only led to frustration for the girl. The next alternative was to produce a computer "mouse" that she could control with her right foot. As only coarse movement was possible a robust joystick-based mouse was designed based on the Microsoft Accessibility features incorporated into Windows 95/98/NT. The design uses a conventional PC keyboard stripped down to the keyboard controller with the accessibility keys externally connected to the joystick microswitches. Full mouse functionality is incorporated including dragging and group operations, which are done using a single press of switch (the drag switching action need not be sustained as the software latches this action ). Figure 2 (a) shows the finished mouse. More information on this device is given on the NRH website. To increase the functionality obtainable with this design a completely button operated version was designed as in Figure 2 (b). This interface in conjunction with a virtual on-screen keyboard such as the WiViK (Bloorview MacMillan centre) allowed the girl to utilise fully all the abilities of modern computers from using specialist educational software to drawing and playing music.

Computer-based Music

As she became increasingly familiar and comfortable with computer use, particularly for drawing and expressing herself creatively, a computerised musical instrument was designed based on the international MIDI (Musical Instrument Digital Interface) standard. This system comprises a software sequencer, a large button twelve-key musical keyboard and a MIDI synthesiser. The large musical keyboard or the button mouse allows the user to input notes to the specially designed sequencer program. This program stores these notes and presents them on-screen as a sequence of simple symbols. The horizontal offset of each dot from the left indicates the time at which the note is to be played and the vertical displacement symbolises the pitch of the note. As an illustration of this Figure 3 shows a screenshot from the sequencer with the representation of a simple melody (Twinkle, Twinkle Little Star) on screen. Once a sequence of notes has been entered, the sequencer can then play back the notes (through the synthesiser) with various speeds, timbres and durations which can be changed on screen. Any MIDI synthesiser can be used as the output instrument including software synthesisers.
Conclusion
It has been shown in this work how the collaborative process between occupational therapists and technologists has helped in the rehabilitation of a young girl suffering from Dystonia Parkinsonism. In particular the use of assistive technology proved invaluable in restoring important occupational performances in the areas of communication, education and artistic expression. We hope to emphasis that an extension, enhancement and enrichment of rehabilitation possibilities can be achieved through this interdisciplinary approach.

Acknowledgements
The authors wish to thank Paul Manning, Michael Collins and Barry Ryan for their help during the course of this study.

References


Bibliography