Letters to the editor

'Guillain-Barre syndrome: a patient's experience'

SIR–Guillain-Barre syndrome (GBS) is the most common cause of an acute or subacute polyneuropathy in children[1]. Patients may show a range of severity, from minimal distal weakness with preserved ambulation to a complete flaccid tetraparesis and respiratory failure. Pain and preserved consciousness are usual in all grades of severity of GBS but may not receive the attention they deserve because of the more acute and demanding medical, respiratory, and autonomic aspects of the condition.

The following personal account of a teenager with acute, severe GBS provides an illuminating and instructive insight into the frustrating, painful, and often frightening effects that are common, and probably under-recognized in this disorder. Although each person with GBS will have a unique experience, it is likely that Mark's experience is shared by many others, including younger children and those who are unable to express their feelings as clearly and eloquently as Mark. It is our hope that this account is read by, or brought to the attention of, all health care professionals who are involved in caring for children and adults with GBS.

'One of my earliest memories on intensive care was lying on my bed unable to move or speak. I knew I was in a hospital but didn't know why. I remember lying there listening to someone's footsteps, faintly at first but gradually getting louder as the person came closer. I thought, It's about time someone came to tell me what's wrong with me and what's going to happen. Then the footsteps faded as the person walked away. It would have been a tremendous help if I was told early on what was happening. Even though I looked as if I was asleep or unconscious, I was perfectly OK on the inside. I do remember my family saying to me that I'll get better, but better from what? Unable to move, speak, breath for myself and also the tremendous pain I was in the whole time day and night—how on earth could I get better from this? Another thing I can remember is how very tired and exhausted I was all the time. I was always trying to move every part of my body, thinking why can't I move?

I had very severe pins and needles all over my body, especially in my hands and feet. It was so bad it was more like daggers and knives than pins and needles. My skin was so sensitive, that the slightest touch felt like salt and vinegar was being rubbed into an open wound. My hips were also giving me a lot of pain, just a dull ache on both sides. Because I had nothing else to do, I used to imagine myself somewhere else doing other things to try and ignore the situation and the pain. I remember when listening to the song 'Eye of the Tiger' on the radio in intensive care, imagining myself in the boxing ring with Frank Bruno going round-for-round with an eventual knock out in the last few seconds of the last round. Victory for me. Suddenly I heard lots of bleeping noises, then they were turned off. I was back in the ring for the heavyweight title when, no longer than the first round, the bleeping noises sounded again! I then realised that the bleeping noises were the machines I was attached to and when I tried to move, my heart rate and blood pressure rose which set the machines bleeping. I was relieved that I had finally found a way of communicating and tried to tense every muscle in my body which in turn set off the machines. I was very frustrated when someone would just turn the noises off. I thought, Why don't they realise what I'm doing .

When I began to move my head I thought, Finally, an end to all the pain because now I can let them know how much pain I was in and the doctors can try and do something about it. Actually, it marked the beginning of a very frustrating time for me. This was because although I could move my head, it wasn't exactly telling them I was in pain. The best way my parents found was that they would go through the alphabet slowly and I would move my head at a certain letter, and by doing this over and over again I could spell out sentences. This was very frustrating because when I got tired my reaction time got slower and I would miss letters and have to go through the whole alphabet again, just for one letter.

Next started the other type of pain: physiotherapy. Every muscle in my body was very tight and had to be stretched, which was very painful. The physiotherapists suggested using gas and air to numb the pain. This caused me to have very frightening dreams during the time I was knocked out and having physiotherapy. I put up with it though because I would have rather had the nightmares than the pain. Chest physiotherapy was also very painful because of my sensitivity to touch. I felt as if my chest was getting punched over and over again even though the physiotherapists were only lightly tapping me. They also used what they called the 'tilt table'. At first glance it looked like some sort of medieval torturing device, which to me it was. The physios strapped me to the table and started to tilt me into the standing position,gradually at first, but then more and more into the upright position. This was my first sight of the intensive care unit and I began to see what the noises were around the room. I was still in great pain and my back felt as if it was being squashed because there was no muscle tone to hold me straight. The doctors tried lots of things to rid me of the pain but everything only worked slightly and nothing ever got rid of all the pain.

Visits from my friends were a great help. I still couldn't speak properly and when I tried to talk it was just a whisper so I don't think they heard me properly and just pretended as if they did because they felt awkward asking me to repeat myself. I was surprised when they came back to see me because I wouldn't have been able to cope with seeing my friends in a situation like that.

I was still unable to move from my neck down and had double vision because my eyes couldn't focus. I remember wishing, If only I could wake up with normal sight, then I could see the television properly to pass the time and take my mind off the pain.

The time came when I was well enough to be moved onto another ward. The only thing I thought about this was that I must be getting better, otherwise they wouldn't have moved me. Physiotherapy was still a big problem to me but there was no easy way of doing it. The problem with hydrotherapy was getting showered and dried. My skin still felt sore and sensitive and the water jets from the shower felt sharp and painful. Getting dried was like having sandpaper rubbed all over severe sunburn. The physiotherapists took

me to the cinema and on trips out which made me feel more comfortable rather than being scared of them. They became more like friends and were always making me laugh when I was in the physiotherapy department which helped me to cope more with the treatment, because I was still in quite a lot of pain.

I was soon moved up to a rehabilitation ward. Again I had become used to the nurses and people on the last ward and didn't want to move again. Things were better on this ward as it was more active and there was more going on. I was a lot stronger and could talk, see and sit up straight. I made lots of friends on the ward because everyone was there for rehab and could talk.

The time came when I could go home for a weekend. This made me very happy but inside, I didn't want to go home for good because for the past nine months the hospital was my home. I had made so many friends, and become used to the routines and daily procedures. I couldn't leave, and in any case I didn't want to. I did want to go back home to my family and friends and my own home and bedroom but my family and friends came to see me so I didn't miss anyone, except our pet dog and the family pets.'

Almost four years after his presentation at 13 years of age, Mark's residual disability is limited to bilateral foot drop (for which he uses ankle-foot orthoses) and slight, but painful, bilateral limitation of hip flexion due to myositis ossificans. Cranial nerve and upper limb function are normal with minimal weakness of hand grip. Muscle stretch reflexes are absent. Mark has applied for a disability licence so that he will be able to drive a car with manual controls.

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'Self-esteem in children with physical disabilities'

SIR–Ten studies that examined the self-concept of children with physical disabilities were evaluated on the basis of age, number of participants, selection of control group, nature and severity of disability, measurement of IQ, outcome of study, and recommendations. These defining criteria were selected because they were considered important in terms of research studies that examine the self-concept of children with physical disabilities.

Three studies found low self-esteem in young people with physical disabilities[1–3]. Seven studies found no significant difference between the self-esteem of young people with physical disabilities and control participants[4–10]. An examination of the methodology used in these studies reveals that an inconsistent picture is being generated in relation to the self-concept of children with physical disabilities.

Firstly, the term 'physical disability' needs careful delineation to address variables such as age at onset of the disability, severity, and the visual appearance of the impairment. This is problematic because of the small samples involved but cannot be overlooked if research conclusions are to be meaningful. Another important issue was the selection criteria of the samples themselves. Harvey and Greenway reported that their sample comprised of 19 children with cerebral palsy (CP), nine with spina bifida, and five with other physical handicaps[1]. Arnold and Chapman report that six of participants in their sample had CP, two had asthma, two were classified as 'delicate', one was said to be suffering from a heart ailment, and two were classified as 'other'[8]. King and colleagues report that their sample comprised of children with CP, spina bifida, cleft lip, or palette[9]. Samples used were often small[2,4,6,8], and many reported wide age ranges[2,3,7,8,10]. While it is acknowledged that it is problematic to find homogeneous samples within disability research, we cannot overlook that attitudes to the self may be specific to the type of disability.

Most studies provided a measure of IQ, however, the reports did not make it apparent which tests or sub-tests were used. The fact that the disabled groups may find engagement with IQ scales problematic because of neurological impairment has mainly been overlooked.

All studies quoted in this research with the exception of King and colleagues[9] made attempts to use a control group. However, it can be argued that it is meaningless to compare the life experiences of young people with physical disabilities with those of non-disabled young people as the life experiences of the two groups are necessarily different due to the presence of the disability. One factor that several of the studies did not take into account[5,8,10] was that some of their sample attended special schooling and some attended mainstream education; indeed, this may have been a confounding factor in the results of these particular studies.

It is not the intention of the authors to ignore the relationship between self-concept and physical disability; indeed it is accepted that research into the self-concept may be a useful preliminary step towards ultimately successful professional intervention to enhance the self-concept of young people with physical disabilities. However, it is argued that researchers need to be open about the methodological problems involved when carrying out disability research.

We should not necessarily assume that the presence of a physical disability by itself is an overriding factor determining the psychological development of children.

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'Analysis of psychomotor development of ten children with Moebius syndrome'

SIR–Moebius syndrome is a congenital non-progressive pathology, characterized by facial paralysis [2,7,8]. Limb defects occur in approximately 50% and learning disability in 10 to 15% of patients [2,6,8,9].

There are reports that children exposed to misoprostol (Cytotec) during the prenatal period, may develop birth defects including Moebius syndrome but a causal direct relationship has not been well established [3,6,9,10,12]. Misoprostol, a synthetic analog of prostaglandin E1, is medically indicated for gastric ulcers treatment. Utilized during the first trimester of pregnancy, it may increase the amplitude and frequency of uterine contractions stimulating hemorrhage and foetus expulsion [3,4,7,8,11,12]. Brazilian women started using this drug as an abortifacient in 1988 although it is an illegal practice in this country [6].

A chart review was conducted to collect and analyze data related to the psychomotor development of 10 children: age range from 1.7 to 7.5 years at the review time; mean age 3.15 years (SD 1.79); seven males and three females. All children were diagnosed as having Moebius syndrome by a geneticist and their mothers reported the use of Misoprostol in the first trimester of pregnancy. These children were followed at Sarah Hospital for the Locomotor System, Salvador, Brazil, by an interdisciplinary team. The geneticist made the diagnosis of Moebius syndrome based on clinical findings. To receive this diagnosis the children presented with at least, uni or bilateral facial palsy (masked face) associated with VI and VII cranial nerve palsy.

Analysis of motor and cognitive development was carried out using development milestones selected from the Sarah protocol for pediatric evaluation. Cognitive evaluation relied on descriptions of cognitive acquisitions according to

Table I: Cognitive acquisitions for children ≤2 years of age

Milestones	*Nr of children showing acquisitions by:*			
	6mo	*12mo*	*18mo*	*24mo*
Response to instrumental sound	3[a]	–	–	–
Look at human face/objects	3[a]	–	–	–
Search a source of sound	2[a]	–	1	–
Follow moving objects	2[a]	1	–	–
Repeat interesting actions discovered	2[a]	1	–	–
Imitate model's behavior directly	–	2[a]	–	–
Explore objects in many different ways	–	1[a]	–[a]	1
Orientation towards a specific objective	–	2[a]	–	–
Find a hidden object	–	1[a]	1	–
Imitate model's behavior indirectly	–	1[a]	–	–

[a]Period expected for achievement of acquisition.

Table II: Cognitive acquisitions for children ≤ 3 years (n=7)

Milestones	*Nr of children showing acquisitions by:*					
	6m	*12m*	*18m*	*24m*	*30m*	*36m*
Response to instrumental sound	6[a]	–	–	–	–	–
Look at human face/objects	3[a]	3	–	–	–	–
Search a source of sound	2[a]	3	1	–	–	–
Follow moving objects	2[a]	4	–	–	–	–
Repeat interesting actions discovered	1[a]	2	3	–	–	–
Imitate model's behavior directly	–	2[a]	–	1	–	–
Explore objects in different ways	–	[a]	1[a]		2	–
Orientation towards a specific objective	–	1[a]	1	1	–	–
Find a hidden object	–	1[a]	–	–	2	–
Imitate model's behavior non directly	–	–	1[a]	–	1	–
Play make-believe	–	–	–	1[a]	–	–

[a]Period expected for achievement of acquisition.

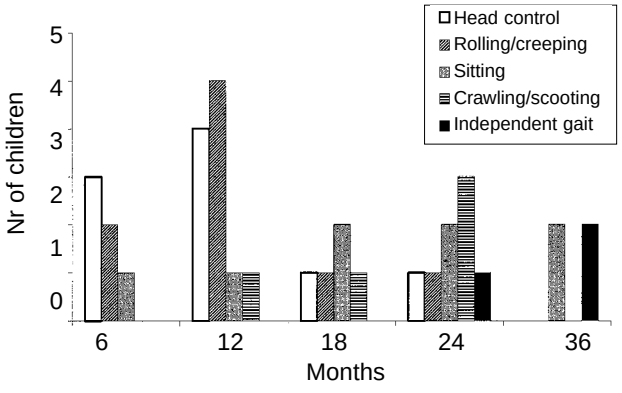

Figure 1: *Motor aquisition milestones according to age*

Piagetian theory. The Denver Development Test was used to compile motor acquisitions data.

All the children showed motor delay although the majority were able to achieve and surpass acquisitions expected for the first semester of life. One child had not yet achieved head control. Eight children had head control and rolled or creeped by 18 months of age. Eight patients achieved sitting. Five patients crawled or scooted. Only three patients had acquired independent walking. Two children walked at around 3 years of age (Figure 1).

Three children were younger than 2 years of age and showed initial cognitive acquisitions (response to instrumental sound, looking at human face/objects, search for source of sound, follow moving objects and repeating interesting actions accidentally discovered[1]). Two of them could imitate model's behavior directly, explore objects in many different ways, orient her/himself towards a specific objective and find a hidden object. Only one could imitate model's behavior indirectly, showing a cognitive development compatible to her/his age (Table I).

Among the seven children older than two years of age, six showed initial acquisitions. Only three children could imitate model's behavior directly, explore objects in many different ways, orient her/himself towards a specific objective, and find a hidden object. Two children could imitate model's behavior indirectly and only a child was able to play make-believe before thirty-six months. One child failed to achieve any of the cognitive milestones (Table II).

Combining both age groups, only two children had developed acquisitions compatible with their chronological age. Behaviour disturbances and presence of stereotyped movements was frequently described (eight children).

The results indicate an important psychomotor development delay in the children studied.

It is important to consider that participants were a highly selected group, referred to a motor disorders specialized center. This may explain the high incidence of developmental delay and orthopaedic deformities (six children had talipes equinovarus).

The retrospective chart review used may have compromised the data in aspects like the reliability of the information obtained from the mothers themselves in relation to the use of Misoprostol.

Prospective studies, using standardized scales to evaluate psychomotor developmen, are necessary to determine the accuracy of the data shown in this study.

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