CASE REPORTS

Rehabilitation after sensory neuronopathy syndrome

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The acute sensory neuronopathy syndrome, first described by Schaumberg et al. in 1979[1], is amongst the rarest of neuropathies, with around a dozen cases reported. It is also amongst the most selective, affecting only the large myelinated sensory fibres of the body (and in some cases the lower trigeminal nerve), leaving patients with intact pain and temperature sensation and motor nerve function but without touch and movement/joint position sense over the trunk and limbs. The responses of individuals with this condition allow insights not only into neurophysiological function but also into the ways in which patients mitigate the effects of such profound and irreversible neurological damage.

CASE HISTORY

Of the case histories of individuals with this neuropathy[2–4], the longest and most detailed is by Cole[5]. More than 25 years ago, at the age of 19, IW lost all sensation of touch and movement or joint position sense after an acute gastro-intestinal infection. He was left with no sensation of touch or body/limb movement or position below the neck, though he had normal feelings of pain and temperature and muscle fatigue, and motor nerve function was normal.

The result was complete incapacitation: without internal feedback of body position he could not make any controlled movement. For six months he required complete nursing for all aspects of daily living. Slowly he realized that by looking at and mentally concentrating on a part of his body he could control its movement. His first meaningful movement after the infection was to raise himself to sitting on a bed. He first had to learn to shift the weight of his head forwards, then move his arms out of the way and then contract the stomach muscles. Without feedback all those movements which we make automatically now had to be made consciously. Once sitting he was so pleased that he relaxed and immediately fell back—any movement had to be concentrated on the whole time, otherwise it would degrade. Over the next two years he was an inpatient at a

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rehabilitation hospital, learning slowly how to feed himself, how to dress, write and then stand and finally walk. All required continuous mental concentration and direct vision of the moving part.

When acquiring motor skills the normal process is to concentrate on them mentally while we are learning and then relegate them to a subconscious motor programme. We may attend to learning to walk or ride a bike, but soon do not need to think about them at all. IW could not develop such programmes and so had to attend to each act each time. Fiercely independent, he preferred a cold meal he had fed himself to the same meal given to him by a nurse. After a year in the hospital he stood and soon after began to walk. Even then, he would fall over if he were to daydream or sneeze while upright—his mental concentration was crucial for mobility. Subsequently, he gained qualifications and worked as a civil servant, living independently on his own and then marrying.

COMMENT

Neurophysiological mechanisms

It is clear that these deafferented individuals use vision as their primary source of feedback. They even sleep with a light on, since if they wake in the dark they cannot move. Visual feedback, however, is slow, and they do move slowly and deliberately. It is also not complete; for instance IW cannot see all parts of himself when walking. This argues for an additional process or processes, which are likely to be the development of elaborate cognitively controlled central motor programmes.

As important as moving in a controlled way is the ability to keep still or maintain a posture. In a previous important study of deafferentation the patient was found to have a motor memory for position of a few seconds[6]. In contrast, IW could keep a given force or position without visual feedback for minutes on end[7]. He, and another affected individual, could reproduce forces between finger and thumb remarkably accurately. They could also discriminate between two weights held in the outstretched arm (with visual inspection) almost as well as controls[8]. These results argue for relearnt central motor programmes run under cognitive control.

It remains unclear whether perception of that central command or its output is necessary, i.e. whether these individuals have developed a central sense of effort or motor efference[9]. I have argued that all the findings can be explained in two ways: either there are sensations of peripheral origin (from small muscle fibres or from vestibular receptors, for instance); or, whilst they have knowledge of the motor command they have ordered for a movement (since they have to order it consciously), they do not have knowledge of that command as it leaves the central

nervous system and so they do not have a perceived efference copy.

IW's ability to order motor commands in a way different from controls was observed in experiments with magnetic stimulation (magstim) to define the focus of motor command[10]. Recordings from surface electrodes over several hand and forearm muscles showed that, when normal individuals moved their thumb, magstim over the contralateral motor cortex led not only to activation of the relevant thumb muscle but also to smaller facilitation of other intrinsic hand muscles. The motor command was not focused only on the relevant muscle but also spilled to surrounding motor nuclei. When IW reproduced this thumb movement only the relevant hand muscle showed a magnetically evoked potential: his motor command was focused exclusively on the relevant motor nuclei. More remarkably, when we asked him to imagine moving the thumb, a similar pattern was seen. His process of imagining led to depolarization of cortical neurons concerned with movement of a single muscle. This is not surprising since for him to order a movement, without feedback from joint or movement sensation or from vision, may be akin to imagining that movement. When we allowed IW to see the twitch movement which resulted from magstim super-imposed in imagined movement, he was surprised—'I did not do that'—since he had ordered only an imagined movement, not a real one.

Cognitive strategies

IW suggests that when relearning to walk he required 100% of his available mental concentration. Now it may be 50% when the ground is even and well lit—an indication that he has been able to delegate part of the programme for walking to a subconscious process. The very fact that he walked at all suggests this, since it is scarcely imaginable that he would be able to monitor all aspects of walking visually, or that all the degrees of freedom of movement necessary to walk could be controlled consciously. One attempt to quantify the cognitive load of walking was to ask IW to press a button in response to an auditory signal while walking. His response time was longer during some part of the gait cycle, suggesting different requirements for cognitive control at different parts of the step[11].

In another experiment IW was asked to make a bread-slicing motion with one hand while sitting on a chair (another motor task) and performing mental arithmetic. This he was able to do so by parcelling his mental energy between tasks (Poizner H, Fookson O, Cole J, unpublished observations). He said he could not have done these tasks while performing another which required precise position-ing—e.g. building a tower of bricks or playing with coins—

since it is motor tasks requiring manipulation or careful positioning of his arm and hand to a position or object which require the most mental concentration.

He describes the mental effort he puts into his movements as being like a daily marathon—a daily marathon of cognition and concentration rather than of simple physical effort. The analogy between neurological disability and sport is apposite. An athlete, however, may train and peak for one or two events a year, while IW lives at his physical and mental limits each day. The only safe place he can relax, knowing that he cannot fall, is in bed, and even to get into bed requires him to be fully awake and thinking.

Disembodied or increased embodiment? Towards a theory of rehabilitation

On a visit to the Johnson Space Center, Houston, IW was watching the astronauts training for a space walk in a giant swimming pool with a space shuttle mock-up sunk into it. He asked how the astronauts learnt to work in such an alien environment. Their trainers replied that they showed each astronaut the task and then invited them to find the best way to do it. They preferred to support and encourage each individual and each crew to find the best way. This was similar to the way IW relearnt to move and walk. The staff at his rehabilitation unit had little precise understanding of his neurological deficit but were expert in giving him the support time and to relearn for himself.

More recently, at a conference on limb bracing, I learnt about the different types of brace: preop, postop, prophylactic, and so on. All had one thing in common: they implied to the patient, 'Your joint is being looked after by something extraneous and so need not involve you.' A strapped-on brace replaced the usual unconscious proprio-ceptive mechanisms to protect the joint's integrity, and the patient was being unconsciously told it was not necessary to attend to the damaged part of his or her body.

Oliver Sacks, in his essay on a woman with a sensory neuronopathy syndrome, called her *The Disembodied Lady*, because of her feelings of isolation from her body once proprioception had been lost[3]. IW has never had such a feeling, except perhaps in the early days when he was unable to move his body usefully. Now he lives a life almost the reverse of this: in order to live and move he has to attend consciously to his body in movement and in repose, in a way previously unimagined. His recovery has depended on substituting his conscious body image as a template to move for the usual unconscious body schema[12]. His recovery has required enhanced, or at least a different, embodiment and sense of ownership and awareness of his body.

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One might consider that such an unusual case has little relevance for more common neurological, medical and surgical disorders. Yet in carpal tunnel syndrome, for instance, sensory loss over the median-nerve supplied skin of the hand may lead patients to say they drop things if they are not looking at them, or that they cannot write properly. They are describing a small area of deafferentation. The solution, if decompression is not successful, might be to educate them to attend to that area more and so be more embodied in it, and to apply cognitive strategies similar to those found useful by IW. After stroke, movement may become more of an effort and require more concentration—a process somewhat similar to that described by IW[13]. A surgical colleague suggested that, in some cases of recurrent dislocation of the shoulder, encouraging the patient to learn to control the muscles of the shoulder better might be useful. More concentration on, and awareness of, the damaged part might, in selected cases, be as effective as reducing personal involvement through splinting.

Although his neuropathy is rare and his solutions to the neurological damage rarer (his motor abilities are in advance of those of other patients with a similar condition), they may contain useful lessons. Rehabilitation from many neurological and other disorders may be helped by allowing and encouraging greater conscious involvement by the patient in control of remaining functions. The remarkable extent and range of IW's recovery points the way cognitive strategies can overcome such deficits.

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Breast cancer after radiotherapy for Hodgkin's disease

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Retrospective studies have shown that, after radical treatment for Hodgkin's disease (HD), the risk of a subsequent solid tumour is heightened[1–8]. Breast carcinoma seems to be a particular hazard when mantle radiotherapy is administered to young women in their teens or early twenties[8,9]. We describe a patient with breast carcinoma following radiotherapy for HD to illustrate difficulties in management of breast tumours and possible implications for the future management of HD.

CASE HISTORY

At the age of 23 a woman received mantle radiotherapy (Figure 1), with 6 MV photons, for mediastinal, clinical stage I A nodular sclerosing HD. The mid-plane dose was 36.33 Gy to the lower mediastinum, 4000 Gy to both axillae and 37.47 Gy to the supraclavicular region in nineteen fractions given by anterior and posterior fields on alternate days.

At 46 years of age, she noticed a mass in the upper central part of the right breast. A wide local excision and axillary sampling was performed and histology revealed a completely excised, 18 mm, grade III invasive ductal carcinoma with an extensive *in situ* component. No vascular invasion was seen and all nine axillary lymph nodes were free of tumour. The

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