Living without Touch and Peripheral Information about Body Position and Movement: Studies with Deafferented Subjects

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Today, as ever, we talk of only five senses: vision, hearing, touch, taste, and smell. This despite the fact that possibly the most crucial of all our senses, position and movement sense or proprioception, was first described nearly 200 years ago. It is so deep within us and so integral to our independence and movement through the world that it has for the most part remained hidden from our personal and collective consciousness.

One reason for this may be that it was impossible to imagine being without it. While the blind and the deaf have been with us, and hence have shown us the consequences of their loss, individuals who have lost the senses of joint position and touch have not been recorded. Recently, however, several such subjects have emerged. This paper will consider some of the scientific work that has been performed with two of these subjects.

In a neurological context, the interest of such patients is that they allow, for the first time, investigation of the mechanism of movement without peripheral feedback in humans. They also allow some reflections on the sense of one's own body and the sense of self in the absence of these important sensory feedback modalities, which, as will be seen, have profound effects on movement and its automaticity.

1   Touch, Proprioception, and the Peripheral Nervous System

That part of the nervous system that lies outside the brain and spinal cord, the peripheral nervous system, may be divided anatomically and functionally into several parts. Those nerves that transmit impulses from the peripheral organs to the spinal cord and brain, the sensory nerves, are conventionally divided into myelinated and unmyelinated nerve fibres. Myelinated nerve fibres are larger and conduct their impulses faster, in part because of their cross-sectional diameter but mainly because of the electrical properties of their myelinated sheath, which acts as an insulator. These large myelinated sensory nerve fibres are divided further into those that originate with

receptors in skin and those with receptors in muscle, the latter called muscle spindles and Golgi tendon organs, which are sensitive to stretch. Smaller myelinated fibres convey impulses that are interpreted as muscle fatigue, tiredness, and some forms of pain and warmth. Unmyelinated nerve fibres convey impulses concerned with pain and cold temperature.

The distributions of the receptors that underlie these sensations and of the nerve fibres that conduct the somatosensory information are not uniform throughout the body. The cutaneous organs are found more frequently in the hand, where they respond to touch and stretch of the skin, while there are far fewer receptors over the back and buttocks. Muscle spindles are found most frequently in those muscles where fine control of movement is important. Perhaps surprisingly, half the spindles in our body are found in the neck muscles, where exquisite control of head position is important for stabilization of the eyes and inner-ear balance organs in space (a fact whose importance will become more obvious later).

The senses of joint position and of movement were first described by Charles Bell in 1833. He talked of the consciousness of muscular exertion as being a sixth sense: "The muscles are from habit so directed with so much precision that we do not know how we stand. . . . If we attempt to walk on a narrow ledge we become subject to apprehension. . . . The actions of the muscles are magnified and demonstrative" (Bell 1979 [1833]). In his original description Bell considered these peripheral organs to occupy a middle ground between the afferent information that reaches consciousness and that which does not. At various times muscular information might be part of one or both of these perceptual sets, which have been described as the unconscious motor schema and the conscious body image (see below).

Phillips (1985) was well aware of the difficulty of talking of a muscular sense of which we are sometimes not aware. He suggested instead that one should talk of the perceptions of movement and of position rather than of the sense of joint position, or proprioception. His definition divorces the impulses originating in given peripheral receptors from a given perception. For example, the muscles concerned with facial expression do not have muscle spindles, and yet there is exquisite sensitivity, mostly at the body-schema level, of position and movement. This information probably arises from peripheral organs sensitive to stretch in the skin. In the hand, information about movement and position of the fingers probably arises from muscle spindles and joint and tendon organs and, most important, from stretch receptors in the skin itself. Muscle spindles and Golgi tendon organs may be more important in joint-position and movement perceptions at the larger, more proximal joints, like the shoulder and hip.

The term 'proprioception' has come to be used, among British physiologists, for those sensory signals that arise from the moving parts of the body, including the head segment and the vestibular apparatus of the inner ear. The visual sense, in contrast, is generally considered as being mainly devoted to exteroception, i.e., to the collection of information about features of the environment in the extrapersonal space. It is possible, however, to talk of visual proprioception as providing visual information about our own body position and movement (see Gibson 1979). The questions of how far visual proprioception may supplement the lack of proprioceptive information arising from the muscles and skin in deafferented patients and how these different sources of peripheral information have different attentional requirements at a conscious level are two of the major themes of this paper.

2   The Physiological Loss in I.W. and G.L.

There have been a number of reports of patients with severe purely sensory, peripheral neuropathies in the last twenty or so years. They were first described in the United States and may have been associated with excess vitamin B_6, or Pyridoxine. These were written up originally by Sterman et al. (1980). Other patients have since been discovered with similar syndromes (e.g., Sanes et al. 1985). The origin of their neuropathy remains still largely unknown, though it is likely that they suffered destruction of their nerves by their own antibodies raised to external infections and cross-reacting to their own nervous tissue. The patients to be discussed here fall into this group. I.W. suffered from a viral diarrhoea and/or infectious mononucleosis, while G.L. had an acute attack of a viral infection. Four years previously she had a classic Guillain-Barre syndrome, a postinfectious neuropathy with severe motor weakness, from which she made a complete recovery.

As a result of these infections both I.W. and G.L. lost sensations of touch and muscular proprioception, I.W. from the collarline down, G.L. from a level at about the mouth. Neither patient is able to perceive touch or movement from below that level. I.W. has apparently normal position sense in the neck, while G.L. has no afferent information from her neck muscles or from the lower part of her mouth and face. Both patients, however, have preserved vestibular information about position and movement of their head in the gravitational field.

Perceptual tests on these patients have been backed up by clinical neurophysiological ones showing a complete absence of large myelinated sensory nerve function. Nerve-conduction studies show no sensory potentials,

and no reflex activity has been elicited. In contrast, nerve-conduction studies of motor fibres and EMG in both subjects is normal. In G.L.'s case, a biopsy from a small sensory nerve in the leg confirmed the complete loss of large myelinated fibres.

Their neuropathies have been extraordinarily selective and complete. All large myelinated sensory nerve function from below their levels has been removed as a consequence of the neuropathy, though motor nerve function and small myelinated and unmyelinated nerve function is apparently intact.

3 Case Histories

I.W. was a successful butcher when at the age of 19 he suffered a flulike illness. He became increasingly weak and at one stage fell in the street and was thought to be drunk. On admission to the hospital he had slurred speech and was unable to feel anything in the mouth, as well as having no touch or positional information from his body. He remembers lying on the bed but being unable to feel it—a floating feeling, which was extremely frightening.

When he tried to move an arm or his trunk, he had absolutely no control over where the moving part ended up, though he had the ability to produce the movement. The facial numbness and problems with chewing disappeared over a few weeks. As the weeks passed, it became evident that little recovery was likely, and he was discharged from the hospital three months later.

He had little understanding as to what had gone wrong but had at least realized that if he was to move with any degree of control, he would need intense mental concentration and constant visual vigilance. After 2 months he had just about managed to sit and was beginning to learn to feed himself.

He spent 5 months at home being cared for by his mother and determined not to admit defeat. He did not allow himself to be seen in this condition, nor did he sit in a wheelchair. He subsequently destroyed all photographs from this time. He taught himself to dress and to feed himself. He preferred a cold meal that he fed to himself to a hot one that someone else would feed him. Then he was admitted to a rehabilitation hospital, where he stayed for the next 18 months.

It was an enormously supportive environment, though it was largely through his own efforts that he stood after a year and began to walk several months after that. He enrolled in the local college and took O (ordinary) level exams successfully (despite severe problems in writing, he neither asked for, nor was given, any extra time to perform the exam) and then

took a job in the civil service. He spent the next fifteen years or so working as a clerk in a job alongside able-bodied people, telling of his bad back rather than having to explain a problem he still did not really understand. After his discharge from Odstock, the rehabilitation hospital, he was not seen by a doctor for about twelve years, during which time he had been promoted at work and married.

The time course and consequences of G.L.'s illness were slightly different. She suffered her neuropathy around seventeen years ago. It affected a higher level, however, since it began at the level of the mouth, extending over the cervical-supplied back of her head. To this day she has no reliable touch or proprioceptive sensation in the tongue or mouth itself and no unconscious ability to control her neck muscles. There were several important consequences of the fact that her neuropathy began at a higher level than I.W.'s. Speaking and chewing were almost impossible, and for several months she existed solely on pureed food. Then she became so exasperated by this that she began to experiment in eating. She learned to push food to one side of the mouth, chew a certain number of times, and then push the food to the back of her mouth and swallow it automatically. Facial expression was severely limited because of the absence of proprioception from the lower face. She refused to look in a mirror for two years after the illness. For a similar time, she had to remember to concentrate to keep her mouth from opening.

She, like I.W., began her own rehabilitation. She was married with a young child and rapidly realized that her main priority was to keep the house together and bring up her son. She decided not to make any concentrated effort at walking but rather settled for a full life from a wheelchair. While I.W. was spending most of his waking life learning to stand and walk, G.L. was immersed in the problems of bringing up a child, doing the housework, and cooking.

Few, if any, of their movements are recognizably normal. Though they show many similarities in movement, the surprising level of walking ability acquired by I.W. compared with the severe impairment in standing and walking observed in G.L. raises an important problem. Psychological problems and differences in their motivational drives are probably not sufficient to explain these striking differences.

The fact that I.W. has intact neck-muscle afferentation, whereas G.L. has not, is most likely the main source of proprioceptive information that allows I.W. to locate his own insentient body with respect to his head segment. This in turn allows him to organize his body balance with respect to the inertial platform of a head oriented with respect to the gravity force field by means of the vestibular apparatus of the inner ear, and with respect

to the stabilized visual field by means of the ocular system. G.L. is devoid of any proprioceptive information about the head-trunk body linkage and therefore is unable to maintain the erect posture of her body that may be necessary for her to achieve, like I.W., an efficient strategy of walking. With her eyes closed, G.L. rotates and bends her head to the right, something of which she is unaware. Likewise with her eyes shut, she is unaware of slow passive rotation of her head. She may lose the location of her head in the same way that she and I.W. lose location of their body segments.

I.W. also has a sense of fatigue in his muscles, whereas G.L. may not, which suggests that I.W. has retained activity in a class of small nerve-fiber sensory afferents in which G.L. has not.

4   Strategies for Everyday Movement

Several factors in these subjects' recovered movement are accessible to scientific investigation to varying degrees. The requirement for visual monitoring of movement is relatively easy to test. On the other hand, these patients' requirement for mental concentration and their dependence on motor programs learned before the onset of their neuropathies are far more difficult to quantify.

Constant visual vigilance is required for any purposeful movement. Both I.W. and G.L. report that they sleep with the light on. I.W. claims that he has to wake up to think about turning over during the night, whereas G.L. reports that she turns spontaneously and changes position while sleeping. She does need light in the room, however, to visually control the actual position of her body in the bed when awakening. Preserved small sensory fibers signaling the lack of blood skin irrigation, which results from prolonged maintained position, may trigger reflex changes of body position in G.L. during sleep, while they apparently awaken I.W.

When one talks with the patients, it soon becomes apparent that this visual feedback can only be used with concentration and intellectual effort. When I.W. first sat up in bed, he was so overwhelmed by this achievement that he stopped thinking about sitting and immediately collapsed. Once he had learned to walk, if he sneezed, and thus disrupted his mental concentration, he would fall over. The limits to how much he can do in a day he describes as having to do with his own mental concentration, rather than the amount of physical effort required.

As well as having temporal limits during a day or a week, I.W. and G.L. also have limits in their focus of concentration. Both I.W. and G.L. use an egg as an example. Having learned the amount of force required for their handgrip to pick up eggs without breaking them (a very challenging

task indeed for both of them!), they can hold them in their hands as long as their attention is not directed toward another task. However, they are sure that if distracted, say during standing for I.W., the concentration required would mean they could no longer think about the eggs and the eggs would be crushed.

We are also forced to use examples rather than measurements to describe their dependence on previously learned movements for the success of motor rehabilitation.

Although both patients claim that they cannot rely on motor automatisms for action and that they have to concentrate on each purposeful movement they want to make, I.W., for instance, is sure, as far as his autonomic locomotion is concerned, that he was greatly helped in rehabilitation by his previously learned movements. In contrast, G.L. stresses the difficulty she encountered in elaborating new strategies for grasping and transporting objects on the basis of visual cues, steering her wheelchair on the basis of thermal cues derived from the cold metallic part of the chair, and even relearning a correct articulation of speech on the basis of her voice sounds.

The more general question of whether or not such patients are able to access a previously learned repertoire of motor habits in the absence of peripheral feedback in order to improve their mobility after the neuropathy is not known and is obviously difficult to investigate. In this connection, how far they would be able to acquire and automatize new motor skills would be worth studying. I.W., for instance, refused to learn to swim, something he had not done before the neuropathy. This, he says, is partly because it would be a completely novel movement but partly because he is inherently unsafe in water, being unable to feel the bottom and unable to see his limbs in order to coordinate movement. He may not have a large enough focus of concentration to move all four limbs together in a coordinated fashion without constant visual feedback. He did, however, pass his driving tests after the neuropathy, with a hand control for braking and acceleration and an automatic gearbox. Driving is one of the most relaxing tasks for I.W. It may not be coincidental that this is the task in which he most closely resembles the nondisabled people.

The distinction between motor activities that are stimulus driven and those that are memory driven (the reactive/projective distinction of Goldberg 1985) is probably apposite here. Stimulus-driven activities generally use ready-to-work motor programs and therefore presuppose the existence of some kind of repertoire of motor habits, motor command programs that can be released and executed automatically. In contrast, memory-driven activities concern goal-directed movements structured on the basis of a

representation of the expected sensory consequences of the planned action once performed. It presupposes the existence of a repertoire of kinesthetic engrams characteristic of the goal to be achieved.

Hence the revival in contemporary studies in motor control of James's old concept of a goal image, which prompts the question of its neural representation. William James stressed the distinction between motor or efferent programs directly (reflexly or automatically) triggered by the stimulus and voluntary actions indirectly driven by way of kinesthetic traces associated with previous movements.

The impairments observed in our deafferented patients raise the basic question of how far reafferent information of proprioceptive origin is necessary to update these traces and/or to steer voluntary action toward its prescribed goal by comparing the desired state represented by these traces and the actual state of our motor apparatus. They also raise the problem of how far proprioceptive information of visual origin may supplement and replace missing proprioceptive information of muscular origin in remapping a repertoire of kinesthetic traces, to restore voluntary control, onto an intact efferent system of motor commands.

To return to James's schema, the apparent persistent absence of stimulus-driven automatisms, often stressed as a motor characteristic of our patients, is at the heart of an unresolved question. Differences between the two subjects in their ability to move deafferented parts, e.g., limbs and body, lead to the conclusion that I.W. may have been far more able to construct motor images. This may have been because, having a stable head and neck posture that G.L. lacks, he was more able to focus his attention on motor planning.

5   Perceptual Frames of Reference

Spatial frameworks are incorporated in our perceptual and motor experiences. The general distinction between a body-centered egocentric frame of reference and a visual exocentric frame of reference is generally accepted (see Paillard 1987). Lacking information about position and movement from their bodies, deafferented subjects should have an impaired egocentric system of reference. Reaching and pointing tasks have been used to test their performance.

Within the calibration process involved in the location of targets in space, several different frames of reference may be distinguished. Visual inputs are coded in a retinal frame, whereas the direction of gaze is egocentrically coded in the body frame, as are most of our motor commands. The object frame is the local space occupied by the object, with its internal geo-

metric relations. The stabilized visual background with its structures and landmarks constitutes a world frame, with respect to which relative positions of the body and of objects might be evaluated.

The relative contribution of these different frames to overall performance may vary according to circumstances. For instance in a normally illuminated structured visual background, the object frame, which may be a target goal for a reaching movement, may give both the visual cues that allow the preshaping of the grip posture to accord with the characteristics of the object and the proprioceptive location cues (derived from the gaze direction of the head and referred to in the egocentric body frame) that allow the directional transport of the hand toward the target. Moreover, a final position adjustment may intervene on the basis of an evaluation of the residual distance between the target and the moving hand in an exocentric system of reference.

Suppressing the structured visual background in a pointing task (for instance by using a luminous target in darkness and by preventing vision of the moving hand) forces the subject to use egocentric cues to perform the task because none are available in the exocentric frame of the visual field. The performance of the deafferented subject (G.L.) is greatly impaired in this experimental condition, whereas that of control subjects is not, which suggests that the latter are normally able to steer their motor activity in either the egocentric or exocentric frame, according to the task requirements, whereas the deafferented patient has a clearly deficient egocentric system. When a world frame is available, however, the patient is as accurate as control subjects in her pointing performance, which suggests that she is able to visually guide her actions in an exocentric frame (Blouin et al. 1993).

Interestingly, when vision of the hand is precluded, preshaping the grip posture to the size and shape of target objects (object frame), which one still finds in control subjects, is absent in deafferented patients. This suggests either that the visual stimulus alone cannot trigger such learned automatisms or that such kinds of learned grip postures are no longer available in these patients. Similar results were described for patients with central deafferentation subsequent to a parietal lesion (Jeannerod et al. 1984). Grip posture may therefore be affected by either central or peripheral deafferentation.

6   The Body-Schema Problem

The divide between body schema and body image is discussed by Shaun Gallagher in this volume. In spite of the clear distinction between the two made by Head and Holmes (1911–1912), confusion persists. Though these

concepts may not always have a clean interface between them (some postural adjustments considered at the schema level can be perceived if concentrated on, for instance), they are very useful models.

Paillard (1980) proposed a similar distinction in extending to somesthesia the functional dichotomy he introduced in the study of visual function between "where" and "what" problems. He suggested that the location of body parts in a body schema (the where problem) is processed differently in the central nervous system from the perceptual identification of the features in a body image (the what problem). Hence, the position of body segments or areas may be either registered as location in a sensory-motor mapping of the body space (that can be reached by a movement of the hand, for instance) or perceived as a position in the perceptual representation of a body image.

Proprioceptive information is necessary for updating the postural body frame (or schema), whereas exteroceptive multimodal information, mainly visual, underpins the central representation and percept of the body image (Paillard 1982).

There are several pieces of evidence showing that deafferented patients may have a preserved body image though they have lost their body schema. G.L., for instance, is normally able to perceive a thermal stimulus delivered to a given point on the surface of the skin of her left arm. When prevented from seeing her body and requested to do so, she is unable to point with her right arm to the place of stimulation. Having lost the ability to proprioceptively update her body schema, she cannot locate that place in her sensorimotor body space. However, she can verbally designate this place in the anatomy of her body ("over my left wrist," for instance), and she even indicates it precisely on a schematic body diagram. In other words, she can locate the stimulus in a perceptual representation of her body, knows *where* the stimulus has been delivered within the frame of her body image, but does not know how to get there in her apparently lost sensorimotor frame (Paillard 1991c).

Of interest here is an opposite syndrome described by Paillard et al. (1983) in a patient with a centrally deafferented forearm following a parietal lesion. This patient, otherwise unable to perceptually detect the presence and content of a tactile stimulus delivered to her deafferented hand in a blindfolded condition, was surprisingly able to point automatically with her intact hand to the place of stimulation. This was described as an equivalent of "blindsight" in the tactile modality. Indeed, in this case peripheral information was still available and processed at a subcortical level, hence allowing the computation of a locality in sensorimotor space, whereas the central process that underpins perceptual awareness of the stimulus was lacking.

Finally, several additional observations concerning the permanence of some basic motor synergies in these deafferented patients are worth mentioning. They concern the preservation in these patients of some inbuilt efferent programs for making anticipatory postural adjustments.

For example, when the subject himself unloads a weight held in his outstretched hand, this produces in control subjects an anticipatory postural adjustment that minimizes upward displacement of the arm, whereas when the experimenter himself unloads the weight, the subject cannot avoid upward displacement (Hugon et al. 1982). If a seated subject is asked to forcibly abduct his leg against resistance, the other leg will also automatically move outward as part of a postural reflex to maintain pelvic alignment (Forget et al., submitted). Similarly, every precision grip involving coordination between thumb and index finger is associated with synergic activity of the extensor of the wrist that compensates for the perturbation by a flexion movement of the index finger (a well-known synergy of Duchenne de Boulogne). EMG recordings have shown that G.L. retains the first (Forget and Lamarre 1990), the second (Forget et al., submitted) and the third (Paillard et al., unpublished results) of these automatisms. (Only the first test has been repeated with I.W. thus far.) This suggests that these postural synergies are not reflexes in origin but are centrally predetermined, either as prewired in the efferent circuitry or as present in a repertoire of inbuilt predispositions, and are preserved for years even when they are of no use.

There are nevertheless other broad categories of anticipatory postural reactions that are clearly under voluntary control and subject to learning: neither subject has any reflex responses to unexpected events, and if I.W. trips, he is liable to fall in a incoordinated way. There are also many adapted feedforward programs that tune motor commands in accordance with the expected dynamic constraints of the planned action (Massion 1984). Muscular proprioception normally plays a predominant role in these adaptive pretuning processes. The problem of how far proprioceptive information of visual origin might allow deafferented patients to compensate for their loss of information about the peripheral state of their motor apparatus awaits further experimentation, yet some evidence on this matter comes from analysis of I.W.'s walking. He relates that walking on flat ground now takes about half the intellectual concentration that it did initially, which suggests some automatization in itself. Analysis of the pattern of activation of muscles during his walking shows that, though very abnormal, there is some phasic activation and relaxation of the calf muscles in relation to the gait cycle that cannot be under visual control and so is presumed to be nonconscious. This suggests that it has become embedded in the motor schema (Burnett et al. 1989).

7   The Problem of Morphokinesis versus Topokinesis

Paillard (1991) has suggested, in the context of drawing and writing, that generating a motor form or orienting an activity within a given spatial frame has different goal requirements, and consequently a different mode of motor commands. Drawing a figure eight in the air, for instance, requires an accurate model of the shape to be drawn (morphokinesis), but this does not need to be located at a specific place in extracorporeal space. Moreover, one can easily change the size of the figure eight without altering the shape. In contrast, reaching to a visual target, say to pick up an object, does require a precise specification of the initial and final locations of the moving limb in an egocentric and exocentric frame of reference (topokinesis).

The deafferented subjects have little difficulty in the morphokinetic mode but have great difficulties with topokinetic movements. G.L. is able to produce a precise ellipse drawn out in the air in front of her. Without vision, the shape of the ellipse remains but the spatial coordinates were altered, as were the orientations of the axes of the ellipse (Teasdale et al. 1993).

There are also differences in their writing. Both can write a legible script, but their techniques for maintaining accuracy with their eyes shut appear to differ. G.L. is very slow and, in making the shapes of the letters, tends to place them in the wrong area of the paper—morphokinesis remains, but topokinesis is degraded. I.W., on the other hand, appears to be more aware than G.L. of the topokinetic problem and eager to achieve the spatial requirements of the task. Thus, with his eyes shut, he moves fast across the page in an attempt to preserve both shape and correct framing of his writing space, and consequently sacrifices some accuracy in making the letters. If he is made to slow down or speed up, the topokinetic component degrades.

8   Production of Force and Corollary Discharge?

In the absence of vision, do these subjects have any knowledge of their motor output? Though there is no disagreement among motor physiologists that perception of movement depends on peripherally originating feedback (and I.W. and G.L. have no perception of movement), is there any evidence that these subjects have some knowledge of force produced? There are several different levels at which this knowledge could arise. There may be some remaining afferent information arising from the periphery, since it is known that I.W. has small muscle afferents intact and both subjects have pain and temperature sensation. Or this could arise purely centrally, either as a perception of the motor command as it leaves the motor apparatus (efference

copy) or as a perception of the command itself as it is generated and before the efferent command is sent (corollary discharge).

A simple way to investigate force production is with the perception of weight. When different weights are concealed within boxes and I.W. and G.L. are asked to discriminate between them with the eyes open, they can discriminate to an accuracy of around 10 g in 150 g. They do this by picking the weight up in their hand and raising and lowering the forearm while looking at it. This perceptual liminal difference is very close to normal. It is thought that they are coding velocity of movement and using visual feedback to see whether or not the movement is smaller or larger, depending on how heavy the weight is. This hypothesis has been validated recently at the University of Laval in I.W. and G.L. using SELSPOT recording of the weighting movement (Fleury et al., submitted).

With eyes shut, I.W. still has an ability to perceive differences in weight, but his liminal difference increases to 200 g in 400 g. The fact that a perception of difference is still present suggests that the origin of this perception is peripheral. This receives support from the fact that when the perceived weights are suspended from the index finger, which is raised and lowered, the ability to distinguish different weights grossly diminishes if the muscles moving that finger are fatigued.

G.L. too demonstrated an ability to produce accurate output of force over a very short period of time. When asked to produce an isometric force by pressing between finger and thumb, G.L. was able to code forces as a percentage of her maximum. She could also give a fairly accurate description of how close the force she had produced was to the force required. This argues for a central perception of force production, though whether it is an efference copy from the motor apparatus or a perceptual copy of the command to the motor centers cannot be determined.

Stimulation of the brain by a powerful short magnetic pulse delivered to the scalp (magnetic motor evoked potentials) can lead to small movements of the hand or fingers. By means of such stimulation it was possible to move I.W.'s finger, while he was maintaining a small isometric force, in a simple twitch movement of which he was not aware. If there is a perceived efference copy, then under these circumstances it does not appear to be at the level of the motor efferent apparatus activated by magnetic stimulation (Cole and Sedgwick 1992).

In experiments with I.W. there is also evidence that over a longer time course he has no ability to perceive and monitor force. He was asked to press down on typewriter keys with each index finger to produce a maximum key movement of 12 cm with a maximum resistance of 500 g. When asked to press two keys down a half, a quarter, or three quarters of the total

distance at the same time or to press one key down and then match that force with a force subsequently on the other key, control subjects were able to do this with relative ease. I.W. found it very difficult either to produce the same force at the same time or to match the force produced by one hand with the other hand. This suggests that he cannot monitor the force he is producing.

There is thus an intriguing difference in I.W.'s inability to maintain forces that he is unaware of and G.L.'s ability to produce calibrated pulses of force quite accurately scaled in a range derived from the production of a series of five maximum forces. The duration of force output may well be crucial here. Both subjects may be able to learn to generate a calibrated pulse of force in an open-loop fashion. However, if the forces have to be altered, they fail. They may have access to some transient corollary signals to regulate the generation of force pulses, whereas information about the sustained maintenance of a given force is lacking.

Additional evidence that there are some interactive visuoproprio-ceptive processes normally associated with motor programs was obtained from experiments in mirror drawing. When asked to trace around a star of David on a table with the index finger with visual feedback through a mirror, and hence inverted, control subjects find it very difficult to draw oblique lines and turn the corners of the star. This is considered to be because of a mismatch between the visual and muscular proprioceptive feedback. It was therefore expected that deafferented subjects would be able to do this task better than controls. Indeed, G.L. was able to do it first time without any conflict (Lajoie et al. 1992). I.W., in contrast, did have a problem at the corners, though he was slightly better than controls.

We may argue that I.W., as compared with G.L., is in general cognitively controlling his motor output far more. While he has neither feedback from the arm nor any perception of his motor program, he does become aware of a mismatch between that program and the feedback he is using to guide his motor output, (just as we are not aware of walking until we trip). He may have elaborated a more secure system of coding motor acts than G.L., whereas she may be dependent almost entirely on visual feedback. He is permanently trying to analyze and understand the motor problem he has to solve (his comments during and after each task extensively illustrate this), whereas G.L. reacts much more spontaneously to every situation.

9   Some Consequences of Deafferentation on One's Approach to the World

Both patients have to give their mental attention to some aspect of movement for nearly all of their waking life. Sitting in a chair requires attention

to avoid falling out of it. On just one occasion in 20 years has I.W. been aware only of what was going on around him rather than actually aware of the process of walking. This has, unfortunately, not become any more frequent. Were he to fall or trip in such a state, however, he would have no awareness of where he was, and this would make the subsequent fall that much worse (when falling, we automatically stretch out our arms; I.W. has no such ability).

I.W. must avoid places which are dangerous to him—dangerous because of the slipperiness of the floor, because of the wind, because of darkness, or because of unpredictability. Crowded places or places where he may not be able to see someone knock him from behind are prohibited. When walking somewhere new, he will map out his next ten yards or so by standing or sitting and studying the area. He progresses by analyzing the way ahead as a mountaineer will work out a pitch up a sheer face of rock. He also has to judge with great care the relation between his size and, say, spaces through which he needs to go. Whereas control subjects may not always attend to this and may rely on reflexes to change direction or stop during a movement, I.W. has to preplan and be alert to the unpredictable.

I.W.'s attentiveness to his surroundings has led him to become in demand by a holiday care service, which uses him to assess hotels for their disability friendliness. He makes sure that a hotel or car park is flat, that the entrances are the right width, that they are protected from wind, etc. His thoroughness is not solely a professional pride but also a personal necessity. For environmental reasons and perhaps because of constitutional differences in the psychological profiles of the two patients, G.L. does not seem to have developed the same alertness as I.W. to potential dangers. Interestingly, when tested in attentional tasks measuring their ability to react to unexpected peripheral cues or to resist distraction, both patients produced astonishing scores that rank them at the highest level of a scale of performance established in well-trained athletes (Nougier et al. 1994), which reflects their need for continual alertness.

10 Body Language

When I.W. and G.L. met recently, it was fascinating to see them together, for they had each independently developed elaborate gestural movements of the forearms during speech. These are possibly some of the most automatic of their repertoire of movements, and are almost entirely morphokinetic. But before I.W. displays this morphokinetic melody, he settles into a posture and calculates how much safe space he has in front of him to gesture within. Then, with minimal visual feedback, he knows how expansive his gestures can be.

They are both well aware of the importance of body language and are strikingly aware that they lost their unconscious ability to communicate through it. Their solution has been to learn a limited repertoire of arm and hand gestures associated with speech that give the impression of an unconscious body language. These movements may not now require constant and complete visual control and are the most automatic of their movements. Both relearned gestural speech under pressure from a social environment that they felt would otherwise complain of their inexpressiveness. To some extent they are using these gestures to deceive others into reacting in terms of an unconscious language: it is an act in more than one sense.

After a decade of living with this neuropathy, I.W. and G.L. have developed a repertoire of gestural movements. It took them several months, however, before they were able to use them in anything like a natural way. G.L. was rather surprised when her attention was drawn to her body language when viewing a video film of herself. She was not really aware of having controlled it at the time. While both subjects' gestures are particularly from the arm and hand, they have also learned to use the fingers to some extent during gestural movements, though not to the same amount as control subjects. They also employ these gestural movements rather more than control subjects.

Both subjects have learned to produce these elaborate movements extraordinarily well. The very fact that they expend so much precious mental energy in learning and reproducing these gestures must reveal something of the importance of expressive body language for us all.

11   Views of Self and of Body Image

One normally considers the body image as being mainly holistic, as being the perception of a gestalt whose components and analytic features are processed largely at an unconscious level. The body image may play a role in organizing and guiding actions as a global percept rather than as a substrate for a sense of local position that allows perceptual awareness of the relations between body parts. It is at the level of the body schema that neural integrations between sensory inputs and motor control translates smooth movement into coordinated movement. Without peripherally originating proprioceptive feedback, I.W. and G.L. have had to use a perceived, visually maintained body image for this purpose. Their use of the body image, and so their perception of it as a concept, may therefore be different from controls' use and perception, since it may operate at two levels: as a holistic gestalt and as a schema replacement.

With this caveat, however, I.W. says that his sense of body image, his sense of his wholeness and configuration, has altered little as a consequence of his deafferentation syndrome. It should be remembered that deafferentation occurred when he was 19. He has little doubt that if the neuropathy had occurred before he had built up an image, then the situation would have been very different. This does not tell us whether the image is built up early in life or it is innate. However, perhaps it is unwise to consider these possibilities as mutually exclusive or to suggest that continued experience does not also play a role. That the body image is built up during early life and then maintained seems unlikely, if only for the simple reason that our size and shape alters during growth. Similarly, our weight and physical prowess may continue to alter during life.

Immediately after the neuropathy I.W. was unable to move, and at that stage, though ill and bewildered, he does remember that he began to think differently about his body: a thing that he could not move had an altered relationship to himself. It was when he began to recover movement that he began to recover a more normal body image, he considers. Though he thinks his body image is normal, he does agree that he finds it difficult to compare with the previous image he had before the neuropathy.

It may be important that G.L. occasionally talks of her body as being a machine on which she imposes commands. It is not clear how literally she feels this. A more accurate description may be that she uses her body as a tool, a passive instrument that can be used to move and to interact with her environment (a machine, though it must be started and controlled, is taken to have some autonomous performance). However, her reduced ability to code accurate movements, because of her additional head and neck deafferentation, does suggest that a normal body image depends to some extent on the ability to move that body. Finally, a tool is not a satisfactory simile in that when we use tools they rapidly become elaborated into a motor schema, and hence are hardly attended to.

I.W. makes the point himself that he has to concentrate more on his body for movement throughout the day than do control subjects. He therefore probably has to be more aware of a body image than do control subjects, and this may have helped him maintain it. Both I.W. and G.L. also have normal senses of pain and temperature. The former was likely very important in avoiding injury, and hence in maintaining a healthy and whole body. Both are useful in determining boundaries. Neither subject has noticed any phantom-limb sensations.

Their body images depend excessively on visual information, and this in itself may suggest it would make such images qualitatively different

if they were maintained with both visual and peripheral proprioceptive information. Much proprioceptive and cutaneous information arises from the hand in a way that visual information could not likely mimic. I.W. agrees that his maintained image might fade in time if not updated and that without vision the remaining senses of pain and temperature, together with fatigue, would be insufficient to keep it. It is not clear, however, that this fading would be qualitatively different for controls.

When talking of boundaries between him and the external world, I.W.'s immediate concern was to stress his need to have a larger personal space surrounding him to avoid the danger of unexpected movements by others. He suggests that, in the absence of touch, his personal boundaries are visually maintained, that he has to keep an idea of the positions of his limbs and their relations to external objects in mind all the time and, by this visual monitoring and visual memory, keep alive both a knowledge of where he is and an awareness of body image. In addition, he needs more space around him to remain safe. On meeting women, he will usually thrust out a hand rather than bestow a kiss upon the cheek. When kissing, he cannot see his body sufficiently to maintain a comfortable posture.

Both patients are very aware of the effects of their neuropathy upon them. The need to concentrate on all movements, and to have developed only a relatively small repertoire of movement has restricted them enormously. The spontaneity of life, the subconscious flow of life, has been removed. When performing any complicated motor act, neither I.W. nor G.L. have sufficient concentration to have their mind elsewhere: I.W. has often said that he cannot walk and daydream at the same time. This focusing of concentration must, one imagines, alter the way in which they view the world and the passage of time. In this, their neuropathies are diseases of consciousness as well as of their peripheral nervous systems. Different motor acts require different amounts of energy: it is easier for I.W. to drive 300 or 400 miles in a car than to stop and refuel for petrol. They view each day in terms of the amount of energy required to move through it rather than for its own sake.

All movement is a form of communication, and both subjects are aware of how abnormal they appear with their impoverished movements. Perhaps this may explain, in part, their extravagant gestural language when seated—extravagant in terms of large and frequent use rather than in the sense of involved or complicated in repertoire. The desire to be seen as normal, or not to be noticed at all, has been an huge motivation, in I.W. particularly.

12   Unavoidable Limitations

In I.W.'s case his success in returning to a nondisabled world for much of his existence has brought many satisfactions. However, because of the extraordinarily rare and difficult nature of his problem and the difficulties in conceptualizing it, this has also led to a lack of awareness in those around him. A brother once said that people stopped asking after I.W. once he exchanged an invalid carriage for a car, assuming that he had recovered.

In children, motor skills, such as learning to write or ride a bicycle, once learned, become unconscious, and so not worthy of praise. We may encourage our children to learn to ride a bike or to swim, but we don't continue to encourage them once they are successful. For I.W. and G.L. to have learned a motor task makes it only slightly easier to perform again. The more they do, the more they are expected to do, and some days I.W. just cannot find the mental effort and concentration to perform all the necesssary motor acts. Something he can do perfectly well one day he just cannot be bothered to do the next, and this he finds very difficult to explain to friends or to get people to understand.

He describes the level of functioning he has chosen to exist at as his peak. Every day he tries to do as best he can, in a way in which some athletes might understand but few others. Athletes in turn are asked to peak a few times in their career or a few times a year; I.W. faces such tasks every day. He is concerned that in the future the pace at which he lives may prove unsustainable. It is to be hoped that the experience of meeting physiologists with an interest in his condition might make him realize the exceptional nature of his response to it and make a possible slackening of his activity more acceptable to him.

Neither subject has managed to use their fingers well since the neuropathy, though G.L. has been able to type. They use one or two in apposition to the thumb in a pincer grip. The fine and skillful movements of the hand are lost irreparably, since they depend on fine motor control under constant tactile and proprioceptive feedback. This suggests that fine, skilled movements of the hand are particularly sensitive to the absence of tactile cues. It cannot have been coincidence that Charles Bell called his book first describing proprioception and active touch, *The Hand: Its Mechanism and Endowments as Evincing Design.*

When the two patients met, there was concern that G.L. would consider herself a relative failure, since she had patently not been able to recover as much useful movement as I.W. In fact, rather the reverse occurred: I.W. wondered whether the unremitting mental concentration required to

reach the level of rehabilitation that he did was worth it when he saw G.L. so well adapted to a wheelchair. Though hardly apparent to outsiders, I.W. maintains that he has had to become selfish to maintain such a level of functioning. Their differing responses to such similar neuropathies shows, among other things, the importance of treating all people as individuals.

13   Conclusions

There are naturally concerns over the extent to which we can extrapolate from these extraordinary subjects, studied repeated over a long time, to others and to controls. However, with appropriate questions and with results interpreted carefully, their study can be important. Their disability and their experiences have allowed an enormous understanding of the role of the sixth sense (see Cole 1995).

Future work may focus on the extent to which visual "proprioception" has taken over from muscular proprioception to provide visually based kinesthetic engrams. The fate of those areas of the brain involved in touch and movement and sensation of position after being deafferented for so long remains to be studied, though there is some evidence for functional reorganization in these areas from experiments on the distribution of visually induced brain waves in I.W. (Barrett et al. 1989). Real-time neural imaging studies may give us answers both to the question of the fate of the deafferented areas and to the perhaps more interesting one of what areas are used to compensate for the massive deafferentation these patients have suffered.

Acknowledgments

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