Trouble in Mind: Stories from a Neuropsychologist's Casebook
Chapter 6: The Mind-Blind Motorcyclist
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Michael was a motorcycle maniac. Like many a 24- year- old male in exuberant health and with a heady zest for life, whenever he could he leapt on his motorcycle and made for the open road. He often rode with one or more of his army mates, but sometimes he rode alone. So on a beautiful day in 1986 it was a stranger who came upon the horrific scene of a motorcycle wrapped around a tree and a crumpled and very still body lying under it. Michael was still alive—just—and was rushed to the critical care unit (CCU) of Auckland Hospital. He was in a deep coma and had multiple fractures where the left side of his body had been crushed by the heavy motorcycle. Even more worrying was the computed tomography (CT) brain scan showing swelling of the right hemisphere of his brain, caused when the right side of his head hit the ground. This dangerous swelling was treated with a drug (mannitol), and his many orthopedic injuries were pinned and set. Within days of his admission, the doctors gave up on trying to save his badly crushed left arm, and it was amputated.

Michael's mother, Joy, almost lived at his bedside, sure that he would know she was there, even although he showed no signs of regaining consciousness. The doctors tried to prepare her for the almost inevitable; that he would very likely die, and if he didn't he would almost certainly be left with severe brain damage and a crippled body from his horrendous orthopedic injuries. Michael's father had died years before and his one older brother lived hundreds of miles away, so Joy was pretty much alone as she struggled to keep up her spirits and her belief that her son would come back to her. Michael's mates and immediate superiors in the army visited for brief periods but could not wait to get out of the CCU, where machines breathed spookily for their silent victims, reminding the shocked observers that accidents do happen, especially to testosterone- fueled young men.

Two weeks after his admission, the doctors were able to tell Joy that Michael would probably survive. The swelling in his brain had subsided and he had made it through the worst. Secretly they thought it would have been better if he had died at the time of the accident; they knew only too well the agony and despair that lay ahead for Joy and her son. But Joy was true to her name, and the day she was told that Michael was out of immediate danger of dying was one of the happiest of her life. The doctors didn't try and curb her spirits; they knew she needed this hope to sustain her over the next weeks, months, and years. A few days later Michael's CCU doctor gently explained to Joy that Michael might not regain consciousness for a long time and she should prepare herself

for the long haul— perhaps just come in and see him for thirty minutes every other day. Joy nodded and thanked him, and continued to spend hours every day at her son's bedside, talking to him and holding his remaining hand. Five weeks after his accident he opened his eyes, and when the relieved doctor asked Michael to squeeze his hand, he did so. With all his tubes removed and breathing on his own, he was transferred to an orthopedic ward, where he remained for another 11 weeks.

Once Michael had left the CCU his head injury was virtually ignored while the surgeons grappled with his severe orthopedic injuries. In an acute hospital where wards and medical services are physically divided according to types of illness or damage, it is often difficult to decide where a patient with multiple injuries should go. In Michael's case he clearly needed extensive orthopedic surgery and specialist care, and as he could by this time talk normally and seemed cheerful and even "chirpy," no doubt the staff were lulled into thinking he had recovered rather well from his head injury. In any case, there was clearly nothing further to be done for his brain either medically or surgically. Even with all the hard work put in by the orthopedic surgeons, the fractures in his left leg resulted in a twisted foot and a severe limp (which would be partially corrected seven years later by foot surgery).

In 1986 the assessment of head- injured patients was very limited, at least in New Zealand. Neuropsychologists were considered an expensive luxury in a stretched public health system, and so it was an occupational therapist who first tested Michael's general functional abilities a month after he had been transferred to the orthopedics ward. Joy had suspected Michael was blind from the time he regained consciousness at five weeks, but the occupational therapist thought that at times he seemed able to look at an object and recognize it. The therapist agreed, however. that his sight was very impaired. A neurosurgeon who was asked to check him over at one point wrote in his notes that Michael had double vision (*diplopia*). This led to a referral to an ophthalmologist ten weeks after the accident, and for the first time Michael's sight was properly assessed. Joy was right all along; her son was blind. But he was *cortically blind*: his loss of sight resulted from damage to the visual cortex of the occipital lobes rather than to his eyes or the optic tracts. It seems almost incomprehensible that Michael's blindness was not picked up by the medical staff for five weeks after he regained consciousness, but it was of some comfort to Joy to know there was nothing they could have done even if they had noticed his blindness earlier.

Strangely, Michael seemed unconcerned by his poor vision as he lay in bed. His optimism and good verbal communication skills perhaps fooled the medical staff into thinking his brain damage had not impaired his cognitive abilities very seriously. Given his right hemisphere swelling on admission to the hospital, it is likely that his lack of

concern or even failure to recognize his blindness was a denial of his problems (called *anosognosia*), a not uncommon finding following right brain damage. On discharge from the hospital 16 weeks after his accident, Michael had mild spasticity of his remaining right hand (his dominant writing hand), and his speech and comprehension of language seemed normal. The New Zealand Foundation for the Blind assessed him as totally blind and enrolled him in their full- time, live- in rehabilitation program.

Twenty- one months after Michael had his accident, he suddenly noticed the moving lights of other cars when he was traveling as a passenger in a car at night. He also began to notice movement on the television. His rehabilitation therapists, excited by this, began an intensive program to improve his vision. Over the next four months, he progressed from being unable to read letters or words to being able to trace the outlines of letters and then recognizing them. At this stage he was able to read short words, such as "it" and "the," but he was completely unable to recognize any objects or people on sight. After a further three months of intensive rehabilitation, he could recognize numbers, use a telephone, and locate his ashtray. He could read simple booklets and his digital watch. When shown a real object or a line drawing or photograph of an object, he could not recognize it, but he could describe the shape of the object and sometimes work out what it was from that description. His ability to recognize objects via touch, sound, or smell was completely normal. For example, if he was shown a bunch of keys, he had no idea what they were, but if the therapist picked them up and jangled them, or as soon as he held them in his hand, he could immediately name them.

His inability to recognize faces (*prosopagnosia*) did not improve, and until she spoke, he never recognized his mother when she visited him. Of course, since people usually speak as soon as they meet, often visitors and staff were unaware that their faces were just a meaningless jumble to Michael. He certainly seemed to have no serious concern about this, and wasn't inclined to tell them. This is likely another aspect of his anosognosia. (Already we can see that anosognosia might be quite a useful disorder to have in these circumstances. Without it, I suspect Michael would have been very depressed and not the cheerful, motivated fellow he was.)

A further three years passed before I entered Michael's contained world. I received a telephone call from his Foundation for the Blind therapist, who had heard about my work with other head- injured patients. She asked for my help in assessing Michael, in the hope that this might suggest new rehabilitation techniques because, as she said, "Something incredible happened to him and he seems to be getting his sight back, but he still can't see!" She also told me that he and his mother were keen to find out more about his problems and would love to be involved in research. Already I was feeling the

excitement: was this that jewel, an eager and bright research participant with a fascinating disorder and a stable brain lesion? I had worked with (and sometimes cried over) too many patients who had terminal brain tumors or neurological disorders that got progressively worse, so hearing that Michael was definitely not in that category was a very big plus.

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The therapist's description of Michael's visual problems— his inability to recognize what he was seeing—sounded like *visual object agnosia*, a disorder that has been known to neurologists for more than 100 years. Numerous accounts of it have been published in the neuropsychological literature since it was described in 1883 by the French neurologist Jean- Martin Charcot. Charcot was in fact the first to describe many neurological and psychiatric disorders, and has been dubbed the "founder of modern neurology." He was a staunch advocate of hypnotism as a valid medical treatment, especially in patients suffering from the then popular diagnosis of hysteria (primarily affecting women, and later dismissed as spurious), but was also well known because of his many famous students, including Sigmund Freud.

Agnosia means "to not know," and thus *visual object agnosia* means "to not know objects by sight in spite of adequate vision." The disorder is "modality specific" in that the patient can recognize the object via the other senses of touch or sound (or sometimes even taste or smell if the object is blue cheese!). It is not a disorder of naming, because patients have no difficulty naming an object they can recognize via a nonvisual sense. The agnosia family encompasses a wide range of "not knowing" impairments, many specific to a particular modality or sense. Thus there are not only visual agnosias, although they are the most common, but also auditory and tactile agnosias. Within each modality there are even disorders that can be discriminated from one another. For example, included within the auditory agnosias—inabilities to recognize sounds in spite of adequate hearing—there are patients who cannot recognize words but can recognize nonverbal sounds and other patients who cannot recognize various characteristics of music but can recognize other sounds, such as a car starting.

There have been descriptions of patients who have visual object agnosia because they cannot perceive shapes, and descriptions of patients who, despite being able to perceive lines and shapes, cannot integrate them into a meaningful whole—the highest form of vision. The neuropsychologist's task is to try to tease apart which type the patient has, as this might be useful information when designing a rehabilitation program, even if only to understand what the patient will be capable of doing. As we shall see, Michael's visual

agnosia was the "highest" type. That is, he could correctly name lines, triangles, squares, and circles but had no idea what a picture of a bicycle was. This type of agnosia is called *integrative agnosia*.

Michael's other strange problem, prosopagnosia, the inability to recognize faces on sight, is often found in the same patients who demonstrate visual object agnosia. In both disorders the damage is to the occipital lobes at the back of the brain. One possible explanation for this co- occurrence is that in some patients, both prosopagnosia and visual object agnosia are consequences of damage to the part of the visual system that represents spatial entities comprised of many interrelated parts. Mild damage to this system would result in a problem in recognizing faces, clearly a difficult task since faces are complex and differ in subtle ways. More severe damage to this system would not only result in prosopagnosia but also cause problems with recognizing complex objects. Because objects tend to be much less complex than faces and easier to distinguish from other objects, more severe damage would be necessary to cause visual object agnosia than to cause prosopagnosia. There are also cases of visual object agnosia associated with *alexia* (an impairment of reading) rather than prosopagnosia. In these cases the problem appears to be in a system that represents numerous discrete parts, such as letters that make up a word and discrete parts that make up an object.

Together, these findings suggest that different systems or pathways in the occipital lobes are concerned with different types of visual representation. Thus patients who appear to have damage in similar areas of the brain may actually have damage to different pathways. In fact, many patients who cannot recognize objects and faces have more damage to the right occipital lobe than to the left occipital lobe, and vice versa for patients who cannot recognize objects and words. This makes sense in that the right hemisphere is more concerned with spatial concepts and the global picture, and the left hemisphere is more concerned with the details that make up the whole rather than the whole itself.

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Today visual object agnosia is considered one of the "classic" neuropsychological disorders, and its study has told us a great deal about the complex higher visual processes we humans use in our everyday lives without so much as a thought. So why did the possibility of studying Michael thrill me? Surely everything that could be discovered already had been? But this is rarely the case in neuropsychology. Given that exactly the same brain structures and neurons are rarely if ever damaged in two humans, it is not surprising that the disorders that result are also unlikely to be identical.

Added to this is the brain itself; no two human brains are exactly alike. Even the brains of identical twins will develop differently according to their life experiences. Thus even if it were ethical for a neuroscientist to cut out a piece of human brain or, using micro-techniques, to cauterize or chemically destroy a specific set of neurons with great precision, it would still not be surprising if the resulting deficits shown by the human guinea pigs were subtly different. So although there had been many well- described studies on patients with visual agnosia before Michael came into my life, I was still excited by what I might find. Indeed, even if all I could find was a classic case of visual agnosia, that too would be fascinating, as Michael was the first person with this disorder that I had ever come across. Reading about these almost unbelievable disorders is one thing, but observing them for yourself is a whole different experience. This is the excitement that motivates postgraduate neuropsychology students and turns their interest in the subject into passion.

But there is a big caution here. Because special case studies like Michael do not come along too often in the working life span of a neuropsychologist, when they do they often become almost "professional subjects." It is obviously important for researchers to guard against exploiting these generous people, an obligation that becomes more difficult over time as a personal bond develops between researcher and participant. The result is that the participant feels reluctant to disappoint the experimenter. Apart from always being alert for early signs of "participant burnout," one way of trying to balance out this potentially exploitative relationship is for the researcher to use the results of the various investigations in an effort to improve the rehabilitation strategies used with the participant. In this way, the neuropsychologist can become a useful member of the rehabilitation team and perhaps contribute to small improvements in the patient's functioning and quality of life. Whether or not this is possible, the majority of research participants gain satisfaction from knowing they have contributed to research that might help others in the future. And last, but perhaps not least, of the benefits to patients of participating in research is the enjoyment they can gain out of doing something different and meeting new people—even scientists! Having a long- term illness or being "disabled" can be very isolating and horribly boring, especially if the individual feels well but is dependent on others. This was definitely part of the attraction for Michael, who had always been and remained a gregarious, chatty, and optimistic soul.

My initial excitement proved to be well- founded. As we will see later in the chapter, my assessments demonstrated that in addition to the classic disorders of visual object agnosia and prosopagnosia, Michael had a significant and very unusual memory impairment and a number of other disorders of higher visual cognition, especially those to do with color, dreams, and imagery.

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Following my telephone conversation with Michael's therapist, I arranged for her to bring him to the psychology clinic at Auckland University. Ten minutes before our scheduled meeting I was having a cup of coffee in the common room when one of my academic colleagues came in. On seeing me he remarked, "I just passed a man in the corridor who, from the look of him, was on his way to see you." When I left the room I saw a tall, well- built young man limping and swaying unsteadily along the corridor, apparently following the person in front of him. His right arm was held at an awkward angle to his body, perhaps in an attempt to steady himself or to protect his body from banging into the wall, and his left arm terminated at his elbow and was finished with a large hook. I greeted his companion (the rehabilitation therapist) and then greeted Michael. He looked for my voice in midair, as blind people tend to do, thrust his right hand out for me to grasp, and with a broad, engaging grin said in a delightful "Kiwi" drawl: "Gidday. Am I pleased to meet you!"

So began a friendship and a research relationship that continued for many years. Michael's eagerness to participate in any new experiment I could think up, and his interest in his own performance and what it could tell us not only about him but also more generally about how the mind works, made him a delight to work with. He was one of my keenest students, and if it had been possible to replace the lost neurons and connections in Michael's brain so that he could function normally again, I would not have been surprised if he decided to take up formal neuropsychological studies.

Of course many neurological patients who have disorders almost as interesting as those demonstrated by Michael do not become long- term "special" cases. Sometimes this is because they are not interested and willing to participate in ongoing experiments, but more often it is because they have other problems that prevent their involvement. For example, the attributes that made Michael an excellent research subject included his youth, normal verbal intelligence, excellent attention span, and ability to concentrate for long periods. These are precious commodities in a neuropsychological subject, as neurological patients frequently have impairments such as poor concentration or difficulty understanding or following instructions, making them difficult to test. Many patients who would otherwise be good neuropsychological subjects tire quickly, but in test sessions with Michael I often tired before him! A final important attribute of his case was the unchanging, stable nature of his brain damage and his neuropsychological problems. Brain scans since his discharge from hospital over a 14- year period demonstrated that no obvious changes occurred in that time. Even after many years of rehabilitation, Michael remained unable to recognize most objects on sight and

remained completely unable to recognize faces on sight. His largely unchanging disorders were disappointing for his rehabilitation but paradoxically valuable for the research. On the bright side—the side Michael invariably took—other rehabilitation strategies were successful: these were strategies to help him live a richer life in spite of his disorders.

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When I first saw Michael, he had undergone a CT brain scan when in critical care but had never had a magnetic resonance imaging (MRI) brain scan. MRI technology was still very new and expensive, and at that time in New Zealand the only way to obtain such a scan was to travel to Australia. I applied for a small research grant specifically for this, and Michael was very excited when the funds were forthcoming. He and his mother boarded the plane for Melbourne and returned after a few days with a beautiful picture of his brain (see Figure 6.1). I had seen many MRI brain scans before, but this was the first I had seen of one of my own patients. The detail was amazing after the CT scans that I was used to. Now, many years later, this scan still looks good to me even though the MRI scans of today are so much better.

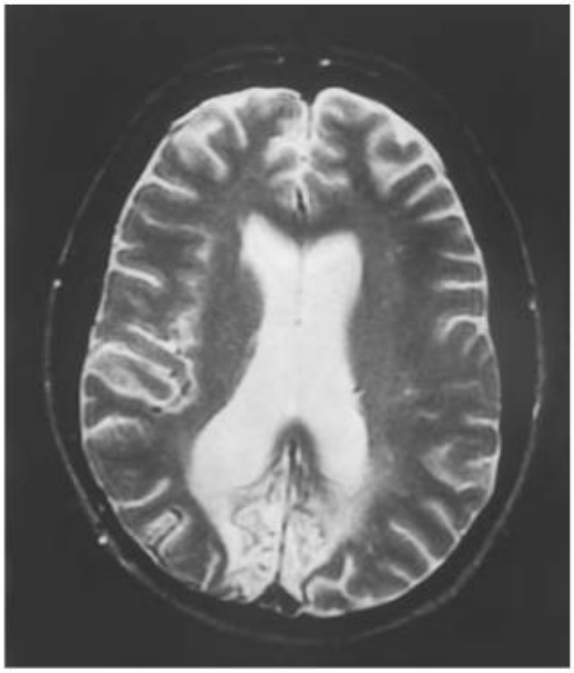

Figure 6.1 The magnetic resonance image of Michael's brain (with the right hemisphere shown on the left side of the scan). It shows areas of damage within the medial aspects of both occipital lobes (lower middle aspect of the scan) affecting gray matter and subcortical white matter, with the damage more prominent on the right.

Look at Michael's scan and imagine it is of a slice made horizontally through the brain above the eyes. The two hemispheres of the brain are like two halves of an apple split through the middle. The *frontal lobes* (one in each hemisphere, above our eyes) are at the top of the picture, and the *occipital lobes*, at the back of the brain, are at the bottom. Between the two are the *parietal lobes*, and if you could see a slice lower down in the brain, near the ears, this area would be the *temporal lobes*, which lie below the parietal lobes. Around the circumference of the brain you can see the intricate folds of the gray matter of the cortex. This is the outside layer of neurons of the brain; on an MRI scan they look like white lines or creases. Michael's cortex looks normal and healthy. Beneath the cortex is subcortical white matter (which looks gray on this MRI scan). The large white symmetrical areas in the middle of the brain are fluid- filled spaces called lateral ventricles. The fluid in these is cerebrospinal fluid (CSF), which is produced at the rate of 600 to 700 milliliters a day within the ventricles, flows out of them through other smaller ventricles lower down in the brain, and circulates around the surface of the brain right down into the spinal cord.

At the lower end of Michael's ventricles are two areas in a slightly splotchy white. This is an altered MRI signal indicating dead brain matter (*infarction*) on the inside, or *medial*, aspects of both occipital lobes, affecting both gray matter (cortex) and subcortical white matter. The area of damage is greater in the right occipital lobe. (Just to confuse things, the brain is usually reversed in CT and MRI brain scans so that the right is shown on the left of the picture.) The lower part of the lateral ventricles (called the *occipital horns*) are larger than they should be because they have extended into the space made when the brain tissue surrounding them died. These extended ventricles suggest that this damage has been there, unchanged, for many years. Other slices of Michael's brain showed that the forward- traveling neural pathways from the occipital lobes to the temporal lobes were damaged on both sides, and on the right side the forward- traveling pathways to the parietal lobe were also damaged. No other areas of significant damage can be seen within the brain.

Like Michael, most patients with visual object agnosia have damage to both occipital lobes—the visual lobes—at the back of the brain, although one lobe might have more damage than the other. The cortex of the occipital lobes is made up of the neurons involved in seeing (primary visual cortex), in visual perception (secondary visual cortex), and in visual knowledge (association cortex). So we would expect patients with visual object agnosia to have damage to the secondary or association cortex, but that at least some of the neurons in the primary cortex would be intact because if the primary visual cortex were completely absent, the person would not be able to see at all.

The next step after obtaining Michael's MRI scan was to assess how well Michael could see. The optometrist's findings were the same as his findings three years earlier: Michael's visual acuity was adequate and correctable to normal with glasses, but his visual fields were markedly reduced, leaving him with five- to eight- degree central fields in both eyes. What this meant is that Michael viewed the world down a tunnel. As long as the objects and pictures he was viewing were held at arm's length and were small, he could scan them and see them in their entirety perfectly well. This important fact allowed his impaired vision to be ruled out as the cause of his more complex, higher visual disorders: He was unable to recognize even the objects and faces he could see at the end of the tunnel. Michael was also found to have no color vision in his right eye and moderately impaired color vision in his left eye.

How his cortical blindness resolved at all almost two years after his head injury remains a mystery. Possibly some neurons in his visual cortex were disabled rather than permanently damaged at the time of his accident, and it took many months for the number of neurons necessary for useful sight to regain their function, perhaps via the formation of new neural connections. Another possible pathway for recovery is the formation of new neurons themselves, or *neurogenesis*. Contrary to what was once believed, we now know that neurogenesis can occur in the vertebrate brain: Rats exposed to an enriched environment both before and after a cerebral insult show enhanced neurogenesis and decreased cell death in the *hippocampus*, a brain area involved in spatial learning and memory. These same rats show improved cognitive performance, especially on tasks tapping those particular abilities. It also appears that the human hippocampal formation produces new neurons throughout adulthood. Although as yet there is no sound evidence that new neurons can form in the human visual cortex, perhaps some degree of neurogenesis was stimulated in Michael's occipital cortex by the intensive program to improve his vision instigated by his rehabilitation therapists when he first began seeing the moving lights of cars many months after his accident. But there is no evidence of any further increase in Michael's visual fields over the past 17 years, suggesting that the recovery in sight that did occur was time- limited.

Now that I had a detailed assessment of Michael's sight and knew where the damage was in his brain, I could try to tie up—or correlate—his behavioral deficits with his specific brain lesions, and I read up on similar cases to see if Michael's deficits and lesions matched them. Already I could see he was a "classic" case of visual object agnosia, and his lesions in the medial occipital lobe matched— roughly at least—the lesions of many other patients with visual agnosia. More fascinating were Michael's other disorders, some of which had not been reported before in connection with these

rather well- defined bilateral medial occipital lesions. The detective in me was eager to get onto the case, and my keen assistant was Michael himself.

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To begin, I needed to know in general how he was functioning intellectually. Of course he could not be assessed on tests that required recognizing pictures and patterns, and his tunnel vision was too bad to permit assessing him on tests that relied on sight. But his performance on verbal tests was quite normal, falling easily within the average range. Even better, because he had been given the same tests three years earlier, I knew that his verbal abilities had remained unchanged over a three- year period, suggesting again that his brain damage was stable. He could repeat seven digits forward and five backward, demonstrating a normal attention span, an important consideration if a person is to be subjected to neuropsychological testing. He scored particularly well on tests of vocabulary and comprehension, a good indication of his pre- accident (or *premorbid*) verbal intelligence.

On tests involving the ability to think abstractly, Michael also performed well. Abstraction is one of the most advanced abilities humans possess, and is usually associated with the frontal lobes. For example, Michael had no difficulty giving the abstract meanings of proverbs such as "A rolling stone gathers no moss." Along with consistent evidence that his performance on other frontal lobe tests and his behaviors in a range of situations in daily life were generally appropriate, his performance suggested that his frontal lobe functions fell within normal limits. This is an important finding in a person who has sustained a severe traumatic brain injury, as frontal lobe damage is very common in this group. Even a CT or MRI scan that shows apparently undamaged frontal lobes is no guarantee that extensive frontal lobe damage has not occurred. Damage that involves shearing and tearing of the white matter tracts connecting the frontal cortex to the rest of the brain is often too diffuse to be visualized. Thus neurobehavioral assessment of frontal lobe dysfunction is essential given the subtle but significant influence of the executive deficits associated with damaged frontal lobes on a range of apparently unrelated abilities.

Michael's speech was fluent and his comprehension normal. He could point accurately on command to lower- and uppercase letters displayed randomly on a page. He could read fluently words in small print held at 1.5 meters to compensate for his tunnel vision. His writing and printing of individual letters, words, and sentences spontaneously and to dictation was slow but generally accurate, and he could read his own printing. His reading of handwriting was slower and less accurate than his reading of print. His ability

to imagine letters was intact. For example, when asked to imagine a letter like a small "c" and decide whether it had curved or straight sides, he always responded correctly.

On tests of verbal memory and new learning, Michael demonstrated moderately severe deficits, suggesting that he had some temporal lobe damage, at least in the left, "verbal" hemisphere. Indeed, his MRI scan showed that the forward- traveling pathways from the occipital to the temporal lobes were damaged on both sides. It is important to note that his verbal memory deficit was not severe, as in the famous case of the global amnesic Henry Molaison, whose story is told in Chapter 7. For example, in contrast to his impaired scores on formal tests of new verbal learning, such as learning lists of unrelated words, Michael demonstrated good memory for conversations that held significance for him. He could recall the gist of conversations he and I had up to three years before and had apparently normal or near- normal recall of the names and other *semantic* (factual) information about people currently in his life. When I telephoned Michael on my return from a year long study leave overseas, on giving him my name he immediately responded by asking me how my trip was and what new tests and rehabilitation ideas I had discovered that might be helpful to him. Many years later I telephoned him "out of the blue" and he immediately recalled who I was. A long conversation about his life since we had last talked ensued, and he had no obvious difficulty with gross verbal recall. For example, within minutes of the beginning of our conversation he told me that Jill, his attendant caregiver, was there with him and that she was the same person who had been his attendant caregiver almost ten years previously, when we had been carrying out our original research. Michael probably also had some impairment of visual, nonverbal learning, but of course this could not be tested because he could not recognize visual patterns—other than simple shapes—or faces.

At last it was time to assess Michael on tests of visual object agnosia. A number of varieties of this disorder have been reported, and it was not a simple task to find out which, if any, fitted Michael best. Generally when assessing a higher cognitive disorder—that is, a disorder involving a deficit of thinking, whether conscious or unconscious—it is important to make sure that the lower levels of the skill are intact. I knew that Michael could see (the most basic sensory level), so I tested his visual perception (the next level in the vision hierarchy). He was accurate at discriminating line drawings of triangles with curved sides from those with straight sides, at pointing to the shortest and longest lines on pages of lines of different lengths, and at pointing to and recognizing different shapes (squares, rectangles, circles, hexagons, and so on). He could discriminate small from large squares and circles, and draw shapes accurately on command. But look at his copies of line drawings of a pig, a bird, and a turtle in Figure 6.2. He made these very slowly, hesitating frequently as he copied them line by line, admitting he had no idea

what he was drawing. When asked what his copy of the turtle was, he said he didn't know, but perhaps it could be a bird. He wouldn't guess the identity of the bird.

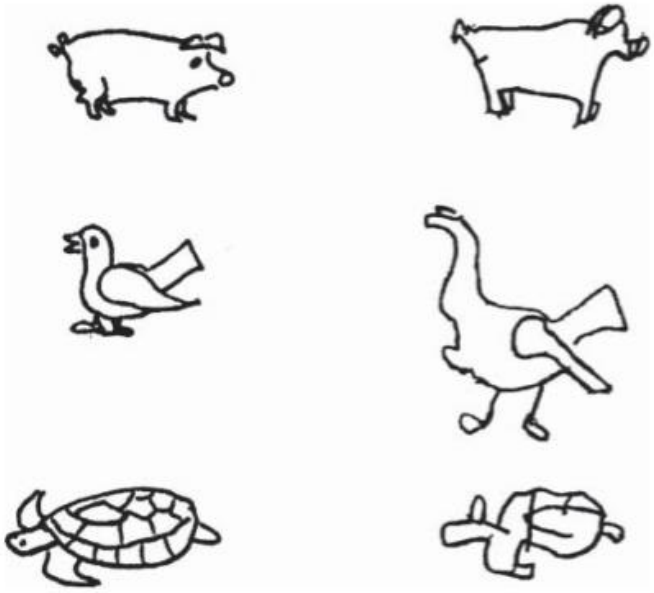

Figure 6.2 Michael's copies of animal drawings showing that he can copy the shape of the animals without knowing what he is copying. The models are in the left column and Michael's copies in the right column.

These tests convinced me that Michael's visual perception was intact. He could not only physically see or *sense* drawings or objects; he could understand them at a basic level of lines and curves and size. But he couldn't put any meaning to what he perceived; he could not tell if the thing was an object or an animal or a scene. Over the years, he did learn to recognize a few objects that he used often, including his ashtray, eating utensils, and scissors. But even then he was very slow, especially if the object was out of context. The way he would go about it would be to describe the shape of the object out loud and then recognize from his verbal description what it was. For example, he would say: "It is a long thin thing and has some thinner bits on one end. Aha, it's a fork!"

On a formal test where he was shown 30 real objects, he could recognize only eight, all of which he had been shown often as part of his rehabilitation program. He would describe each object to himself and then make a guess for every object whether or not he got them right. A yellow feather he called a "flower," a safety pin he called a "clothes peg," and a vegetable peeler he called a "razor." When shown a key, he described it thus: "A circle; there is a long, thin piece off one side; it is smooth on the top but seems to have a jagged edge on the bottom." But he could not recognize it. As soon as he picked it up, however, he recognized it instantly as a key. This was true for all the test objects. If the object could make a sound, he recognized it, and if he could feel it he

knew what it was. His ability to recognize photographed objects, realistic three-dimensional paintings of objects, and line drawings of objects appeared slightly more impaired than his ability to recognize real objects. For example, he recognized a real telephone in four seconds (he often used a telephone), but he was still unable to recognize a line drawing of a similar telephone after 35 seconds of trying. I then said "ring, ring," and he said "Oh, is that what it is, a telephone!" One hour later, shown the same drawing, he was able to identify it correctly in five seconds.

He recognized two items, a house (eight seconds) and a spoon (six seconds), in a series of 20 black- and- white photographs of common items. He was shown 30 realistic colored paintings of common objects, animals, and people in context (e.g., sheep in a paddock, a baby eating from a bowl, apples on a tree, fruit in a bowl) taken from a series of books used for teaching words to three- year- old children. He was asked to name specific objects in each painting and was able to name only two items correctly, a pen and a person, and even those took him three and four seconds, respectively. He was unable to say whether the person was male or female, adult or child. He could identify some animals as animals after describing their form to himself, but he was not able to identify any specific animal correctly. In all the above experiments, he was given a minimum of 60 seconds—a very long time—before being permitted to give up, and longer if he thought he might be able to recognize the item given even more time.

Michael was also severely impaired in naming line drawings of single objects or living things. Of a series of 60 items, he named only three. Again, with repeated exposures over a short period he became faster at recognizing his own descriptions, but this did not indicate a true improvement in his recognition, as he soon forgot what his own verbal description was most likely to describe. For example, when first shown a line drawing of an elephant, he attempted to describe its shape but was still unable to recognize it after 30 seconds. He was then told it was an animal, but this did not assist him. He was then given the names of five animals and asked which one it was. He correctly selected an elephant and was able to point to the trunk. One hour later, he was shown the same drawing, and after nine seconds he was able to identify it as an animal "because it has four legs," but he could not be more specific. When shown the same line drawing two years later, he was once again completely unable to identify it or even correctly categorize it as an animal.

I gave the same 30 real objects used in the visual recognition experiment to Michael to manipulate with his right hand, his left arm having been amputated. I put large objects, such as a telephone, on a table in front of a blindfolded Michael, and placed his right hand on the object. Small objects like a key were placed in his right hand. He was free to

manipulate the objects as he wished, and when necessary they were held steady while he explored them. He named all 30 objects correctly within one to four seconds.

While blindfolded, Michael was asked to name 20 sounds, such as water being poured from a jug into a cup, the rattle of a bunch of keys, the sound of rain outside the building, and scissors cutting paper. He named all sounds correctly within one second.

Just to be sure Michael didn't have a problem with knowing what an object or animal was in a more general sense (that is, not just via its appearance), he was given the name (e.g., "cat") or other identifying information (e.g., "something that goes 'meow' ") about an object or an animal and asked to describe it. His performance was normal in almost all cases. When given the names, he was able to describe the function of the 30 objects used in the visual and tactile recognition experiments (e.g., he could say what a spade is used for). He was also able to describe living things and natural phenomena (e.g., trees, specific animals, clouds, mountains) and provide factual information about them. For example he said a cow provided milk and meat, a tree was a large plant that grew in the ground and provided shade on a hot day, and a cloud was a fluffy thing that floated in the sky and sometimes produced rain. In contrast, he was frequently unable to describe accurately the shape of the object or living thing. For example, when asked what a cup was, he said it was made out of pottery or china and was for drinking. When asked about its shape, he said that it was hollow with a handle to hold it. When asked if he could visualize a cup, he said he could not, but that he knew he picked it up by a handle to drink from, and it must be hollow to hold coffee. When asked what a canary was, he said it was a small bird that whistled, but he could not quite see its shape. He thought it had two legs. He did not know what color it was, but he thought it might be blue.

Michael could quickly and accurately draw from memory triangles, squares, rectangles, crosses, and circles. He could draw on command shapes of different sizes and lines of different lengths. For example, he responded correctly when asked to draw a line across the top of the page and underneath it a line of half the length of the first one. He could draw a large circle above a cross and a small circle below the cross. He could also draw on command some objects with simple shapes (e.g., a rugby ball, a basketball, a rugby field, an apple, a banana). He could draw in a simplistic fashion some more complex common objects on command. For example, he was able to draw a recognizable house and key (see Figure 6.3), but his drawing of a flower looked more like a palm tree and he drew a bed as a rectangle with four legs. He would not even attempt more complex or unfamiliar objects, such as a telephone or a broom, because he said he could not think how they might look. He was also unable to draw or visualize any specific objects, such as the house where he grew up.

Figure 6.3 Michael's drawings of a key and a house from long- term memory. These show that Michael can draw in a formulaic, nondetailed manner some very common objects.

When asked to visualize and describe a surfing beach where he had spent a great deal of time before his accident, and which he had been to since, he said he could imagine the waves and the sand and hear the surf, but he did not describe the dramatic scenic surrounds of bushes, cliffs, and rocky peninsulas. When asked the color of the sand, he said it was pale, when in fact it is of a distinctive black iron color.

It appeared, therefore, that Michael was unable to visualize or imagine objects and scenes from his long- term memory store, or, alternatively, that he had lost the visual memory "templates" (patterns of specific objects) themselves. The drawings he was able to do and his verbal descriptions of visual forms from memory were impoverished and nonspecific and probably represented stylized prototypes of general object categories. Michael also said he didn't have dreams, perhaps suggesting a loss of the ability to generate visual images.

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Michael had a total inability to recognize any faces on sight, and this has not improved over the 24 years since his accident. Practice and familiarity did not help; he never recognized his mother until she spoke although he saw her almost daily. He was unable to pick out any familiar face, including his own, in family photographs taken before and after his accident. I showed him a collage of photographs of faces of people famous and well known to him before his accident and faces of people he would never have seen. When asked to point to any faces that seemed more familiar to him, he said that none seemed familiar. When asked to describe verbally from memory the very distinctive face

of a famous New Zealand prime minister who had been in the public eye for many years before and after Michael's accident, Michael said (accurately) that he had a lopsided smile. When he was then asked to guess which of the faces on the collage was this man's very distinctive face, he pointed to a man whose face he had never seen before.

Michael's ability to discriminate gender, age, and expression of faces was also greatly impaired. He would sometimes guess gender correctly by the length of hair, but when this was controlled for, he performed at chance levels. When asked to say whether a face looked happy or sad, he would rely on the shape of the mouth and made numerous errors. When asked how old the person in a photograph of a baby might be, he said it looked like an old man because it did not have much hair. When he was shown photographs of single faces and asked to match each one to the identical face in a pair of faces, he performed correctly on all eight trials, but took a long time, generally matching the face by the hairline.

Michael also had a problem with identifying colors, whether in isolation or in the context of an object. He commented that he saw everything as shades between white and black. When asked to name the color of tokens, he was correct only on white and black ones. All other colors he named either "white" or "pale," or "black" or "dark." Yellow was "white," and dark blue was "black." When asked to group tokens according to their color, he made two piles; one he called "dark," the other "pale." This loss of color perception is termed *achromatopsia*, and it results from bilateral lesions in the *prestriate visual cortex* extending to the temporal lobe, lesions that fit well with Michael's. When Michael was asked to give the colors of named (but not seen) objects, animals, or natural phenomena, he was usually correct with regard to natural phenomena that are frequently and stereotypically associated with their color names, such as blue sky, white clouds, blue sea, green grass, and white snow. He made numerous errors on most objects and living things. For example, he guessed a banana to be either green or blue, and although he described a strawberry as a small, sweet berry that grows on low bushes and is eaten with sugar and cream, he could not visualize its shape or its color. Toast was "dark" and an apple was "brown." He described a sparrow correctly as a small, common bird, but he was unable to visualize or remember its color. He guessed it to be blue, and when told it was brown, he said this did not enable him to visualize it. When asked what color his skin was, he first said it was blue; told that was wrong, he said pinkish white. This loss of the ability to visualize colors would probably impede his ability to remember or recall colors.

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Michael's loss of color memory was trivial compared with another of his memory problems. In apparent contrast to his reasonable verbal recall of life events since his accident, Michael had a striking loss of personal memories that extended back from his accident into his early childhood. His mother, other family members, and friends he knew in school and in the armed forces spent hours retelling Michael about his past. Michael would comment that he could not really remember a described episode, although he sometimes remembered it from having been told about it since. A striking example of this loss of all personal memories prior to his accident—*retrograde autobiographical* memory loss—was his forgetting of his 21st birthday, just three years before his accident. This was a large celebration remembered very clearly by his family and friends.

On a test of autobiographical memory where the individual is asked to describe facts (such as the address of the house lived in as a child) and personal events (such as an incident that occurred in primary school), Michael's score was well into the abnormally low range, more so for personal events and for events further back in time from his accident. He did retain some auditory memories from before his accident. While still a patient in the hospital he could recognize the horns of specific ships in the harbor close by; he had previously spent three years in the navy specializing in sonar. He was also able to recognize the voices of his friends, and even today he can still remember both the melody and words of some songs popular prior to his accident.

His severe loss of memory for personal events from before his accident is in dramatic contrast to his memory for personal events since his accident. Of course even his current memories don't contain any visual information. For example, he described a barbecue he had been to four days earlier thus: "I had a ball. It was a beautiful, sunny day after all that rain we've been having. They had some really good spicy sausages, and of course I got drunk later in the evening." He said he could recapture the sounds and tastes of the occasion in his mind and the good feelings he had, but he had no visual images or memories of the occasion whatsoever. Michael's memory loss for events prior to his accident is hard to explain when we look at his MRI scan. Occipital lesions do not result in amnesia, and although some pathways to the temporal ("memory") lobes were damaged, he had no obvious damage to the temporal lobes themselves. The only way to make sense of this deficit is to postulate that his autobiographical retrograde amnesia results from his inability to recall the visual components and visual images associated with his personal memories. Given that humans are generally very reliant on the visual aspects of their experiences, perhaps Michael's inability to recall or imagine visual aspects of objects, faces, and colors impoverishes his memories so much that it is as if he doesn't have any recall of an event. For example, even if Michael's recollection of his 21st birthday party could be activated by hearing a recording of a speech made at

the party or by the music played, his reconstruction of the event would be so constrained by not being able to remember the dominant visual aspects such as peoples' faces, the room the party was held in, or his birthday cake that he might not recall the event at all.

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Michael's inability to recognize objects or faces is, in a sense, almost more debilitating than being totally blind. He is always striving to recognize what he sees, which can act as a barrier to learning how to cope without sight. Because he can read it is not necessary for him to learn braille; yet his tunnel vision makes reading extremely tiring. His problems in moving about due to his loss of one arm and his old orthopedic injuries are only exacerbated by trying to avoid objects that appear to loom up at the end of a tunnel and that he is not able to recognize. However, while he sometimes comments on his inability to make mental pictures and his loss of dreams, these impairments do not seem to worry him.

His mildly impaired ability to retain new information makes rehabilitation difficult at times. For example, learning how to cook with one arm and an inability to visually recognize a saucepan, carrot, or tomato requires that everything in the kitchen be kept consistently in the same place. Michael forgets what goes where and has difficulty remembering to follow simple but important safety measures when cooking. He cannot go out alone, because he cannot learn new routes as many blind people can and because his poor physical mobility makes it too dangerous.

His loss of pre- accident autobiographical memories does not seem to upset him unduly, perhaps because his mother and friends have spent many hours telling him about his past to help him gradually build up some sense of where he comes from. He remembers his old friends well (from their voices), but he cannot recall anything they did together. It is a credit to Michael's friendly and happy nature and great sense of fun that he has retained some good friends over the years since his accident. They still collect him and take him to their homes for a meal or party, where he enjoys drinking and socializing.

Given his many disabilities, it would be understandable if Michael often felt depressed and frustrated or became disenchanted with the ongoing grind of rehabilitation. Remarkably, this has not happened, and Michael consistently maintains his positive outlook. When asked how he does this, he replies that he is lucky to be alive and he could be much worse off. As he says, he has regained his sight, and his ability to walk about is improving, especially with the latest operation on his foot. He does not appear

to harbor any underlying feelings of bitterness or anger about his fate, although he does, of course, feel depressed at times about the future. If he were always happy, it would indicate that he had poor insight into his problems and suggest that he had sustained some frontal lobe damage. Throughout his rehabilitation, Michael has generally maintained a high level of motivation, although he becomes frustrated with the slow pace of establishing the sequential steps needed to cope safely with activities of daily living.

After living in the Institute for the Blind, in his mother's home with a full- time caregiver and rehabilitation therapist, in a good friend's home, and in a home for young disabled people, six years after his accident Michael achieved, at least in part, his desire for independence. He moved into a rental apartment where he lived alone, but with an attendant caregiver, Jill, coming in daily to help him and accompany him when he wanted to go out. He learned his way around the rooms and became skilled at using the telephone and microwave oven, washing dishes, and cleaning. Jill has continued to come to his house for a few hours every weekday for the past 17 or more years. She is now a grandmother and she and her family have become Michael's close friends. As Michael joked, "Jill and I are like an old married couple, but without the sex!"

Some years after his accident Michael received from the New Zealand Government Accident Compensation scheme some belated compensation, which, with the help of Jill, he invested wisely for his future, but which also enabled him to follow some of his dreams. He purchased his own house and in 1999 he flew to the United States with Lou, a friend he made following his accident. Lou worked for an organization called Bikers' Rights and assisted motorcyclists injured in accidents. He and Michael became good friends, and Lou would often take Michael for trips in the sidecar of his Harley-Davidson. Michael has always loved motorcycles, and the accident that almost took his life and certainly changed it forever has not in any way diminished his passion. His total amnesia for the accident itself and his very hazy memories of the many months of operations and pain that followed was probably a significant factor in allowing his love of riding motorcycles to remain untarnished.

Michael had a dream to ride Route 66 in the US, and Lou said, "Why not?" Michael's mother was initially worried that he would end up in another accident, this time perhaps a fatal one. But his mother joined the dream- believers after thinking about Jill's wise comment that Michael could sit in his big leather chair in his safe house for the rest of his life, or he could follow his dream, even if the risks were great. After much organization and planning, Michael and Lou flew to Los Angeles from New Zealand, then to San Antonio, where they picked up a Harley- Davidson. They then proceeded to ride, with Michael in the sidecar, to New Orleans, Memphis— including Graceland—and

Chicago, where they picked up Route 66 and rode to Los Angeles, Las Vegas, and back to San Antonio. In Chicago they fell off their motorcycle and Michael suffered a bad gash to his left leg, which had been severely fractured in his original accident. The Chicago surgeon who attended him was a motorcyclist himself, and stitched him in such a way that he could continue his journey across the States. As Michael told me later, "My leg was a mess when we got back to New Zealand, and I had to have more surgery, but man, it was worth it!"

When they returned, Lou made up a photograph album of their travels, but then realized that this wasn't much use to Michael. He then had the wonderful idea to put together a series of musical CDs to remind Michael of their trip. For example, the first CD includes "Leaving on a Jet Plane," "New Orleans," "Memphis, Tennessee," "Carolina Blues," "Miami Vice," "The Painted Desert," "Needles and Pins," "Grand Canyon," "Viva Las Vegas," "Tombstone," "Riders on the Storm," "Waltz Across Texas," and "Harley-Davidson Blues." Michael believes that he can remember parts of his trip, especially when cued by the songs on his CDs, which remind him of those locations or the music they enjoyed on their trip. Even if his memories of details of the trip are vague and nonspecific, the emotions and impressions of that trip—the feel and smell of the motorcycle and his leather gear, the wind on his face, the perfumes in the air, the music (often also linked to his past before he had the accident), and simply riding Route 66—are instantly and vividly brought to life for him by the music and of course by sharing stories with Lou, Jill, and his mother. With friends like Lou and Jill, who needs professional rehabilitation therapists! Michael's story is truly one of triumph over adversity, with a little help from friends.

Michael thinks he can "see" a little better now, and he and Jill sometimes even go to movies—although he admits that it is the soundtrack rather than the visuals that allows him to follow the plot. He thinks that he can recall autobiographical memories laid down since his accident that involve sound, tactile sensations, and emotions. Certainly, his spontaneous recall is clear for many of the tasks we did during the original assessments many years ago.

Michael's story not only teaches us about some fascinating neuropsychological disorders but also provides lessons in courage, stamina, determination, and an all-important ability to laugh at oneself when all else fails. These characteristics have enabled Michael to progress to a reasonably independent life despite minimal recovery of his visual and memory impairments. Now 48 years old, he has not been able to visually recognize his mother, friends, or the world around him for 24 years, half his life span. Yet he remains the positive, effervescent person I first met and continues to live life to the full. As he likes to say, "Well I can see a little, but can conceive of all!"

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For many clinical neuropsychologists, doctors, nurses, and rehabilitation therapists who work with patients like Michael, the theoretical insights provided by research studies are interesting but must take a backseat to the more urgent need to assist the patient to a state where he or she can regain a reasonable quality of life. To do so requires practical knowledge that often can be learned only by working with brain- damaged patients. Experienced rehabilitation therapists know, almost by intuition, when a patient can be pushed a little further and when it is time for a rest or a change of activity. They learn how to predict and prevent the sudden outbreaks of aggression that can happen to any patient as a result of frustration and fatigue, and they know when a touch of humor will lighten the situation and help the patient to laugh at himself or herself. They know when a patient needs to cry and to express anger or helplessness, and they learn how to listen to what the patient needs to help him or her at these times.

The rehabilitation therapist must have an abundance of practical knowledge, patience, stamina, determination compassion, and humor. The rewards come from working with a person like Michael, who in spite of massive disabilities courageously continues to make small positive steps—and occasionally giant strides—while retaining his good humor and endearing himself to all who have the good fortune to come within his auditory or tactile orbit.

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