23

Neuropsychology

- The modern theory of MODULARITY sees some psychological functions such as sensory processing and language as LOCALIZED in particular parts of the cortex, whereas other functions such as intelligence are diffusely organized throughout the cortex.
- The left hemisphere is DOMINANT for language processing in 95% of right-handers and 65% of left-handers. The right hemisphere is specialized principally for visuo-spatial processing.
- LICHTHEIM'S MODEL of APHASIA can explain both BROCA'S and WERNICKE'S aphasias, as well as the DISCONNECTION SYNDROMES of CONDUCTION APHASIA and TRANS-CORTICAL APHASIA.
- The deficits found in Broca's and Wernicke's aphasia may not merely be of speech production and perception, but perhaps of deeper linguistic functions such as syntax and semantics.
- Cases of ACQUIRED DYSLEXIA suggest that normal reading is carried out through two separate processes: a PHONOLOGICAL ROUTE, which uses GRAPHEME-PHONEME TRANSLATION, and a semantic or LEXICOGRAPHIC ROUTE, which can read irregularly spelt words and has access to meanings as well as sounds.
- Right hemisphere lesions typically produce the AGNOSIAS, in which sensation and perception are disconnected, and CONSTRUCTIONAL APRAXIA, in which there is an impaired understanding of three-dimensional space.

Selective damage to the brain, by disease or trauma, can result in psychological deficits which illuminate both brain structure and psychological processes. To be successful, NEUROPSYCHOLOGY has several requirements: a method for locating cerebral lesions; a technique for relating psychological deficits to anatomical deficits; and an adequate theory of normal functioning (mere 'natural history' of syndromes is not sufficient to give a clear understanding of *normal* processing).

The localization of lesions has improved dramatically in the past two decades, before which brain slicing at post-mortem was the only reliable technique. Techniques such as COMPUTERIZED TOMOGRAPHY (CT), and MAGNETIC RESONANCE IMAGING (MRI), can now distinguish abnormal areas in the living brain. Newer techniques, such as POSITRON-EMISSION TOMOGRAPHY (PET) show cerebral metabolism and hence functioning,

which is physiology rather than anatomy.

If a patient has a particular brain lesion and a specific functional deficit, does that mean the function is carried out at that site? Surprisingly, the answer is No. The lesion might cause a general deficit (say, in memory or cognition) and the task affected is merely the most vulnerable to the general deficit (a threshold effect). The fallacy can be seen in the apocryphal story of the foolish scientist who amputates the legs of a cockroach, and then, after noticing that the cockroach no longer jumps at loud sounds, concludes that the animal hears through its legs. Psychological functions can only be localized by DOUBLE DISSOCIATION (the previous situation being single dissociation). If patient 1 has a lesion at site X and function A impaired (but B intact), and patient 2 has a lesion at site Y and function B impaired (but A intact) then assuming the brains are organized in the same way, then *either* function A is localized at X *or* function B is localized at Y (but with only two patients, we do not know which). *What* is localized must be interpreted with care. Remember the old anecdote of removing a valve from a radio set, hearing it start to whistle, and falsely assuming the valve's function is to stop the set whistling. Pathological functioning can only be interpreted through an adequate knowledge of *normal* functioning.

Some functional processes may not even be localizable. Since the mid nineteenth century there has been debate between two separate schools of neuropsychology. The LOCALIZATIONISTS, inspired by the successful localization of sensory and motor cortex, and by lesions that specifically damaged language (see below), argued that each psychological ability had its own cerebral location. The alternative view, of MASS ACTION, was put most forcefully by the American psychologist Karl Lashley (1890–1958) in the 1930s; the location of a lesion was unimportant, and what mattered was the *size* of the lesion, larger lesions producing greater deficits. Experiments in rats had shown that the deficit in learning a complex maze related only to the *amount* of cortex damaged and not its location; and in humans a decreased IQ related only to lesion size and not location. These two schools have recently been reconciled in the concept of MODULARITY: input-output processing, concerned with sensory analysis, motor control, and language, is organized as discrete, functionally autonomous MODULES which can be individually impaired by localized lesions, whereas general intellectual activity is diffusely organized, and therefore only affected quantitatively by lesions.

Lesions can have different effects upon the brain. DESTRUCTION of an area completely impairs its functions. DISCONNECTION of the fibres between two areas allows each to function normally but a specific deficit occurs if one area needs to communicate with the other; very specific testing may be needed to detect the problem. ISOLATION occurs when all the connections to and from an area are damaged, leaving

the area functioning but unable to communicate: the lesion is functionally equivalent to destruction but there is no actual anatomical damage to the area. Lesions also affect more distant parts of the brain: cerebral shock, or DIASCHISIS, probably due to loss of excitation, can inhibit remote areas and physical effects, such as CEREBRAL OEDEMA, a local swelling of damaged tissue, can compromise blood supplies to other areas. RECOVERY can take place in several ways, perhaps due to lesions being incomplete, or to functional take-over by unaffected areas.

An additional complication in interpreting brain lesions is that although the left and right cerebral hemispheres are almost identical anatomically, functionally they are very different. In mid-nineteenth century France, Marc Dax and Paul Broca (1824–1880) noticed that patients who had lost their speech after cerebral damage had *right*-sided hemiplegias, and hence language must be in the *left* cerebral hemisphere, which was referred to as the DOMINANT or MAJOR HEMISPHERE. The situation was soon seen to be more complicated, with language dominance related to right- and left-handedness, although left-handers were *not* the mirror-image of right-handers; in 95% of right-handers the left hemisphere is dominant, whereas in left-handers, only 65% show left-hemisphere dominance. A majority of both right- and left-handers therefore show left hemisphere dominance, but left-handers show greater variation than right-handers. Left-handedness and language dominance are probably inherited by the same single Mendelian gene which produces FLUCTUATING ASYMMETRY, a random determination of handedness and language dominance, which means that neither right nor left-handers 'breed true', and that monozygotic twins are often discordant for handedness.

Cerebral dominance can be assessed by several techniques. In DICHOTIC LISTENING a subject listens simultaneously through headphones to separate strings of words in each ear, and then recalls the words, those presented to the right ear being remembered better, the RIGHT-EAR ADVANTAGE (REA) reflecting left hemisphere dominance. The technique is useful for assessing dominance in populations, but cannot ascertain dominance in an individual with certainty. TACHISTOSCOPIC VISUAL HALF-FIELD TECHNIQUES require a subject to stare at a fixation spot, and words are then presented briefly to right or left visual half-fields; a right-visual field (RVF) effect demonstrates left hemisphere dominance for language processing, but again cannot be used reliably for assessing individual dominance. Reliable assessments are sometimes necessary before neurosurgery (as when a central brain structure should be approached through the non-dominant rather than the dominant hemisphere, in order to avoid the disaster of post-operative language loss). An effective technique is INTRA-CAROTID SODIUM AMYTAL INJECTION (the WADA TECHNIQUE) in which sodium amytal is injected directly into an internal carotid artery, producing general

anaesthesia of the cerebral hemisphere on the same side for three or four minutes, the other hemisphere remaining fully conscious. If speech impairment occurs after the injection then that side is dominant for language.

Dichotic listening and visual half-field studies also show that the right (or so-called 'non-dominant' hemisphere) is superior to the left at processing non-verbal stimuli, such as musical tunes and rhythms, pictures and faces, and it is now accepted that the hemispheres are COMPLEMENTARY, the left preferentially processing material that is verbal, logical, analytic and spread out sequentially in time (as in speech), whereas the right hemisphere processes information in a holistic or intuitive manner, the separate components being analysed simultaneously and in parallel (as in pictures). This functional separation is clearly seen in the syndromes occurring after brain lesions, and which form the rest of this chapter. For simplicity all patients will be assumed to show the archetypal pattern of left-hemisphere dominance for language-related behaviours, and right hemisphere dominance for non-linguistic abilities.

Neuropsychology was founded by Broca when he described a series of patients with what is now known as BROCA'S APHASIA (synonyms: ANTERIOR, NON-FLUENT or MOTOR APHASIA). Aphasia is a specific deficit of language processing, and in Broca's aphasia there is an inability to *produce* spoken language (although perception is unimpaired, the patient can understand commands, and tongue and larynx control is normal; patients can sometimes even sing the very same words of songs that they are unable to speak). The speech that is produced is painfully slow, often being disconnected grunts and noises that individually are similar to those of speech but without integration. Less severe cases show TELEGRAPHIC SPEECH, consisting mainly of nouns, with few verbs or prepositions. Prosody, or intonation, is unimpaired. Classically the lesion occurs in the third left frontal convolution, just anterior to the sensorimotor areas controlling lips, tongue and face. Broca's aphasia is contrasted with WERNICKE'S APHASIA (synonyms: POSTERIOR, FLUENT or SENSORY APHASIA), described two decades later, in which speech is produced fluently, and sounds superficially normal although produced in such excess as to be called an 'intolerable oquacity'). Detailed speech analysis reveals many pronunciation rrors, 'table' being said as 'bable' (a PARAPHONIA), or being compounded ith a related word, such as 'chair', to produce 'chable' (a PARAPHASIA). hese patients seem unable accurately to monitor their own speech; nilar disruption of pronunciation can be produced in normal dividuals with DELAYED AUDITORY FEEDBACK, a half second delay between eaking and actually hearing one's own voice through headphones. The relationship between Broca's and Wernicke's aphasia is well n in the theoretical model proposed by Lichtheim in 1885 (Fig. .1). Sound enters via the ear and is processed firstly by the auditory

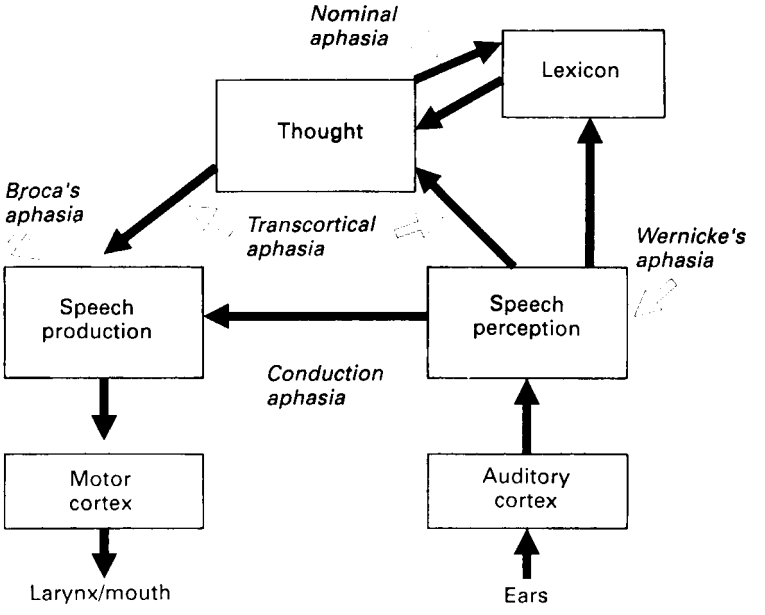

Fig. 23.1 Lichtheim's model of the relationship between speech perception, cognition and speech production. The lexicon is not a part of Lichtheim's original formulation. Open headed arrows indicate the site of lesions producing specific syndromes.

cortex and then by Wernicke's area, which translates speech into the internal code used for 'thought'. The output of Wernicke's area passes to a 'central processor', which processes the information, 'thinks' about it, and sends an output message to Broca's area, which translates the message back from internal code into a speech based code, and then sends its output to the motor cortex and thence to mouth and larynx. In addition, there is a direct link between Wernicke's and Broca's area, which does not require translation into the internal code of thought. Lichtheim's model not only accounts for the deficits of Broca's and Wernicke's aphasias, with their destruction of specific cortical areas, but also explains two DISCONNECTION SYNDROMES. In CONDUCTION APHASIA, patients have only a single, very specific deficit; they are unable to repeat exactly a sentence that is spoken to them. They can understand what is said, they can speak normally, and can speak about the message, even paraphrasing it (e.g. 'The cat sat on the mat' becomes 'the kitten sits on the carpet'), but are simply unable to repeat messages verbatim. The direct route between Wernicke's and Broca's areas is damaged, meaning that an input cannot be copied exactly from input to output and instead must be translated into the internal code and then its meaning translated back into speech, losing accuracy in the process. The lesion is in the ARCUATE FASCICULUS, the bundle of white matter connecting Broca's and Wer-

nicke's areas. The converse syndrome is TRANSCORTICAL APHASIA, which is associated with diffuse brain damage, often due to carbon monoxide poisoning, in which the cognitive processing portions of the brain are disconnected from Broca's and Wernicke's areas; inputs to Wernicke's area pass directly to Broca's area without cognitive processing, so the patient can *only* repeat sentences, without evidence of comprehension, and produces no spontaneous utterances. The only exception to strict repetition is an alteration of grammar, so that sentences refer directly to the speaker, 'How are you today?' becoming 'How am I today?', implying that grammatical analysis occurs in either Broca's or Wernicke's areas.

A further syndrome, ANOMIC APHASIA or ANOMIA, was not described by Lichtheim but can be included in his model. The major defect is in remembering names of objects and people, and it can be regarded as being an amnesia for semantic (not episodic) information. Although superficially normal, patients use circumlocutions to avoid difficult words, often using the words inappropriately (*e.g.* 'I have three children, a son and two females'), and they fail completely when asked directly to name specific objects. The deficit is entirely of *conscious retrieval* from the mental dictionary or LEXICON, since words heard in speech are recognized and interpreted, and the correct names of objects can be recognized as correct.

Although Wernicke's and Broca's aphasias seem to be deficits of speech perception and production respectively, the linguist Roman Jakobson (1896–1982) suggested the deficits may be broader, Broca's aphasia being a deficit of sequencing or syntax, and Wernicke's aphasia a deficit of comprehension or semantics. That interpretation is supported by experiments showing that Broca's aphasics misunderstand the perception of syntax in *spoken* speech (AGRAMMATISM), confusing commands such as 'Put your hand on the book' and 'Put the book on your hand', which differ in syntax; typically the patient guesses at the more likely or more reasonable meaning. Since word order is particularly important in grammar, the speech problem may also be a problem of ordering or *sequencing*, the subtle movements of larynx and mouth in speech not being made at precisely the correct time.

Aphasia is considered in depth because it is common, occurring frequently after cerebrovascular haemorrhage or ischaemia. It must however be emphasized that the most frequent presentation is as GLOBAL APHASIA, with deficits of both speech production and perception. Despite its serious implications for the patient, it is of relatively little neuropsychological interest.

One of neuropsychology's major successes is in discovering and understanding problems of reading after brain-damage in adults (ACQUIRED DYSLEXIA which must not be confused with DEVELOPMENTAL DYSLEXIA, that occurs in childhood and may require very different types

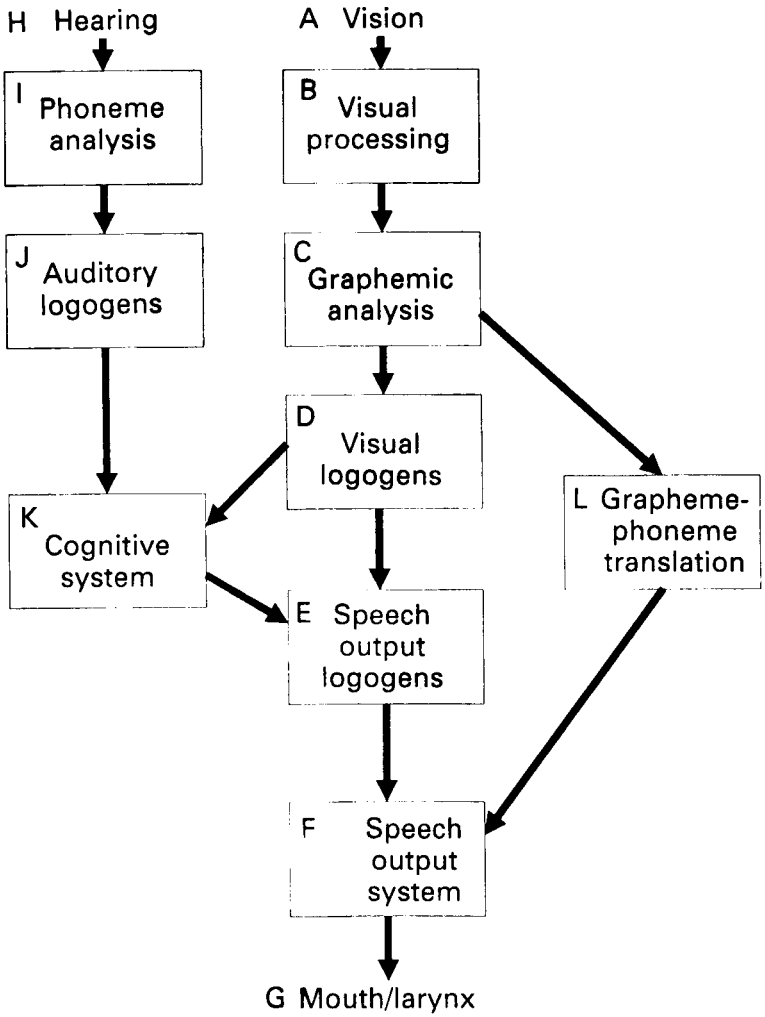

Fig. 23.2 A model of the centres involved in reading words aloud. See text for further details.

of explanation). The dyslexias are rare, but tell us much about normal functioning. Although superficially straightforward, reading is a complex skill (as any illiterate adult will tell you). A visual analysis must first distinguish curves, horizontal and vertical lines, etc. (stage B in Fig. 23.2), before GRAPHEME recognition (stage C in Fig. 23.2) — graphemes being the letters, symbols, etc. of writing; 'A', '*A*', 'A' and 'a' all being the same grapheme. Individual graphemes are then combined with other knowledge (such as context, expectations, the visual shape of the word, etc.) by the system known as the VISUAL INPUT LOGOGEN (D). The logogen outputs are understood as meanings by the cognitive system (K), and a speech output can be produced by

the SPEECH OUTPUT LOGOGEN (E) that converts meanings into the phonemes of spoken speech, which are then sent to the equivalent of Broca's area (F). In addition direct routes between input and output (as in Lichtheim's model of aphasia) allow simple repetition of material, either by a direct link between D and E, or by a grapheme-phoneme translation system (L), which from any particular grapheme combination produces the standard sound pattern. Such a sequence of analyses would be required by any system processing writing, so that a reading and speaking computer would have to use a similar method. However, it is supported in man by patients with specific deficit syndromes.

The simplest form of acquired dyslexia is LETTER-BY-LETTER READING (also known as PURE ALEXIA and ALEXIA WITHOUT AGRAPHIA), in which reading is painfully slow, individual letters being read slowly aloud, one at a time, and then the word recognized only after it has been spelled out ('H, O, U, S, E, house'). Since these patients do not suffer from AGRAPHIA they can therefore write but cannot read what they may just have written. They are lacking access to the stage of graphemic analysis (a lesion of stage C in Fig. 23.2). Letters are therefore recognized only as arbitrary shapes, so that after the lesion the sound-shape associations have to be relearnt. The route of reading is therefore A-B-Memory-G-Spoken sound-H-I-J-K-E-F-G. A and K therefore only connect via the medium of spoken sounds produced in the outside world. Reading is characterized by slow recognition time, which is a linear function of word length.

PHONOLOGICAL DYSLEXIA is frequently not recognized by patients themselves, since the deficit is in reading aloud NON-WORDS (such as 'bif' or 'kaj', for which most individuals can produce sounds with ease). The deficit is in the direct 'grapheme-phoneme' route (box L); graphemes cannot be converted to sounds unless a logogen already exists for the word. One of the normal reading routes, A-B-C-L-F-G is therefore deficient.

DEEP DYSLECTICS are like phonological dyslectics in being unable to read non-words. However, in addition, they cannot read abstract words lacking in imagability (e.g. 'virtue' or 'sophistication' rather than 'house' or 'curlew'), and find function words (such as verbs and prepositions) more difficult than content words (nouns and adjectives), so that 'ambulance' can be read, but not 'am', or 'bee' can be read but not 'be'. Occasionally there are also semantic errors, so that 'ape' will be read as 'monkey'. This syndrome is the dyslectic equivalent of conduction aphasia, without direct routes between visual logogens and speech output logogens, or a grapheme-phoneme route, so that all information passes through the cognitive system, where meaning is extracted but specifically linguistic information is lost, so that words with diffuse meanings (such as 'be') are lost or distorted. Non-words, which have no meanings, simply cannot be read at all. All reading

is therefore through the route A-B-C-D-K-E-F-G, the normal routes D-E and C-L-F being damaged.

SURFACE DYSLEXIA is the inverse of phonological dyslexia, reading being satisfactory for non-words, function words and abstract words, but impaired for longer words (which do not particularly affect the deep dyslectic) and showing an effect of REGULARITY OF SPELLING (which is not shown in deep dyslexia). As any foreigner will affirm, English spelling is eccentric, some words being spelt and pronounced in a regular way, whereas others are ORTHOGRAPHICALLY IRREGULAR, so that, for instance, 'gauge' as normally pronounced should be written 'gage' in regular spelling, or should be pronounced 'gorge' if its spelling were actually regular (as in the regularly spelt 'gaunt'). Surface dyslectics read 'gauge' as 'gorge' (and 'broad' as 'brode' and 'trough' as 'true'). The defect in surface dyslexia is not entirely clear, the difficulty with longer words being similar to that in letter-by-letter reading, but surface dyslectics have the additional problem with irregular words, which suggests that the visual input logogen is working only partially, so that many regular spellings are available but access to the lexicon for unusual and irregular spellings has been lost. The most likely explanation is that the route D-E is damaged, so that reading is either through the routes A-B-C-K-E-F-G, or A-B-C-L-F-G.

Acquired dyslexias often involve the ANGULAR GYRUS, at the junction of the temporal, occipital and parietal areas, a site well placed for auditory-visual integration. Nevertheless, lesions can involve many cortical sites, and in deep dyslexia the lesions are so extensive in the left hemisphere, that it has been hypothesized we are observing the pure reading of the non-dominant right hemisphere.

The left-hemisphere syndromes of aphasia and dyslexia have been described in depth because they illustrate well the methods of neuropsychology. The remaining syndromes will be treated more cursorily, being included principally to illustrate the wide range of neuropsychological deficits.

Two other symptoms are typically associated with the left hemisphere, both involved in motor control. AGRAPHIA, an inability to write, can occur in association with or independently of aphasia and dyslexia, and is classically associated with lesions of EXNER'S AREA, in the frontal lobe adjacent to Broca's area. A more general deficit of motor control is APRAXIA, in which praxis (or action) is impaired at a high intellectual level. Despite no weakness, spasticity, or evidence of upper or motor neurone lesions, apractics are unable to organize movements into a meaningful sequence, as in striking a match or combing hair. Lesions are often at the left angular gyrus, which may store kinaesthetic engrams (or memories); damage not only impairs movements with *either* hand, but also causes difficulty in perceiving and understanding movements made by other people. SYMPATHETIC APRAXIA occurs only in

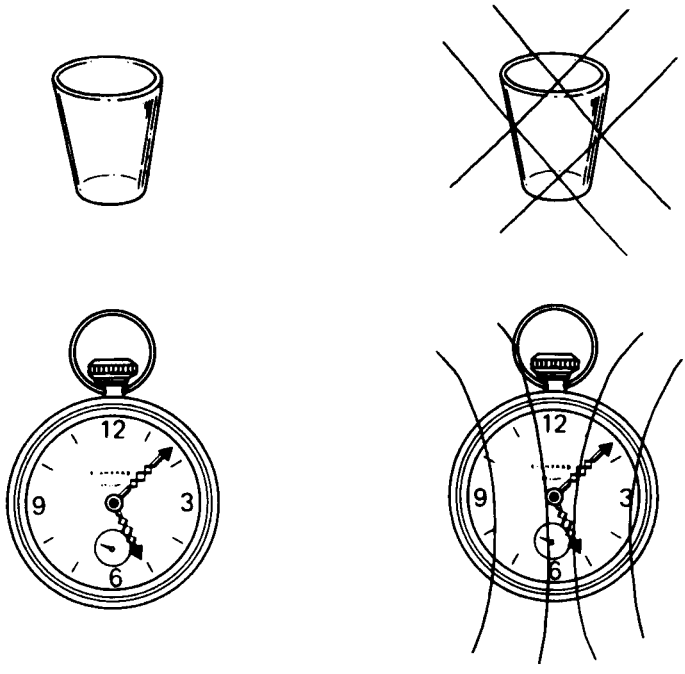

Fig. 23.3 Visual agnosics find it difficult to recognize the objects on the right hand side, in which crossing lines have disrupted the overall outline. The original test was devised by Luria. Reproduced from Kolb B and Whishaw I Q (1985), *Fundamentals of human neuropsychology*, 2nd edn, New York, W H Freeman, 212.

the non-dominant hand, and is a disconnection syndrome, the non-dominant motor cortex having lost contact with the kinaesthetic engrams stored in the dominant angular gyrus, due sometimes to a tiny lesion in the body of the corpus callosum.

The syndromes of the non-dominant hemisphere are well-recognized and described, but have been studied far less, in part because of the greater difficulty of using non-verbal stimuli in experiments.

The AGNOSIAS are syndromes in which sensory processing is complete, but information is not interpreted to form a meaningful whole; SENSATION is present without PERCEPTION (see Chapter 2). The syndromes are modality specific (visual agnosia, auditory agnosia, tactile agnosia, etc.). In visual agnosia, despite being able to identify lines, curves, colours, etc., patients cannot recognize objects, particularly if, as in Fig. 23.3, additional lines confuse the overall outline. Agnosias are often very precise; for instance COLOUR AGNOSIA is an inability to recognize appropriate colours for objects, due to a disconnection of the visual cortex from areas concerned with the memory of colours (Fig. 23.4). PROSOPAGNOSIA, a specific inability to recognize familiar faces, is usually regarded as a right-hemisphere syndrome (and

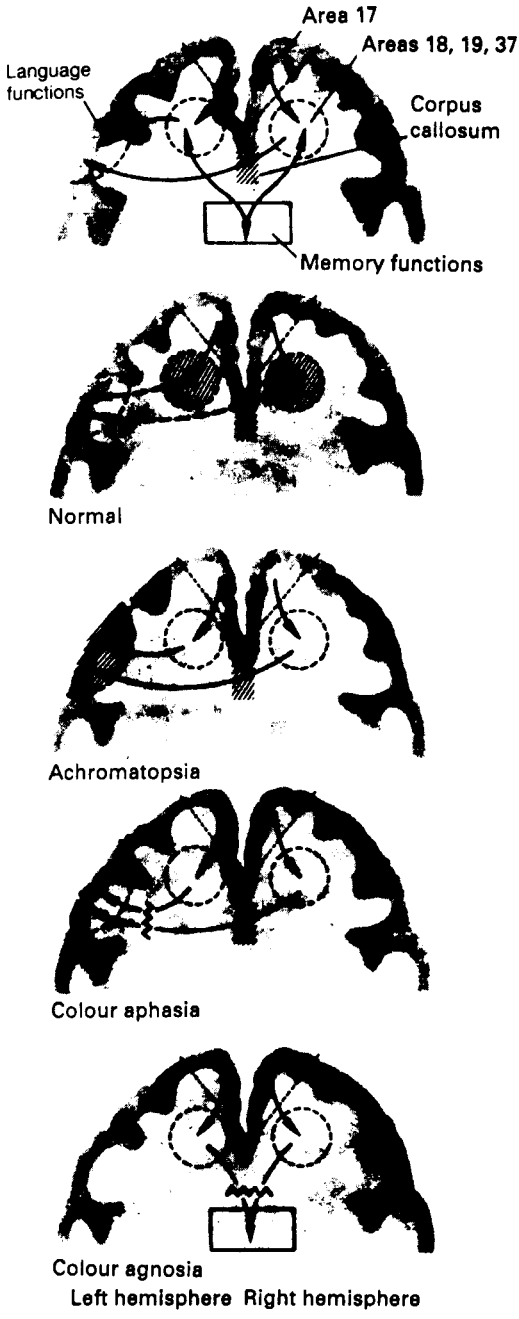

Fig. 23.4 A model for deficits in colour perception. In the normal individual the retinal image is processed by area 17 (primary visual cortex), and then by areas 18, 19 and 37, which access memories of colour information (shown by the square box), before transmitting information about the name of the colour to the language functions of the left hemisphere. ACHROMATOPSIA is a form of cortical colour-blindness, in which colours cannot be distinguished from one another due to bilateral destruction of areas 18, 19 and 37. COLOUR APHASICS show normal differentiation of colours, but are unable to name them, due to disconnection of the language areas from areas concerned with colour perception. In COLOUR AGNOSIA there is normal discrimination of colours but an inability to recognize appropriate associations of colours with objects (e.g. of red with tomatoes) due to disconnection of colour perception areas from memory. Reproduced with permission from Kolb B and Whishaw I Q (1985), *Fundamentals of human neuropsychology*, 2nd edn, New York, W H Freeman, 212. Copyright 1980, 1985 by W H Freeman and Company.

visual half-field studies show a right hemisphere advantage for face recognition), but it is now accepted that bilateral lesions are present in most cases, accounting for the rarity of the syndrome. In UNILATERAL NEGLECT, the patient ignores half of the visual world, simply denying

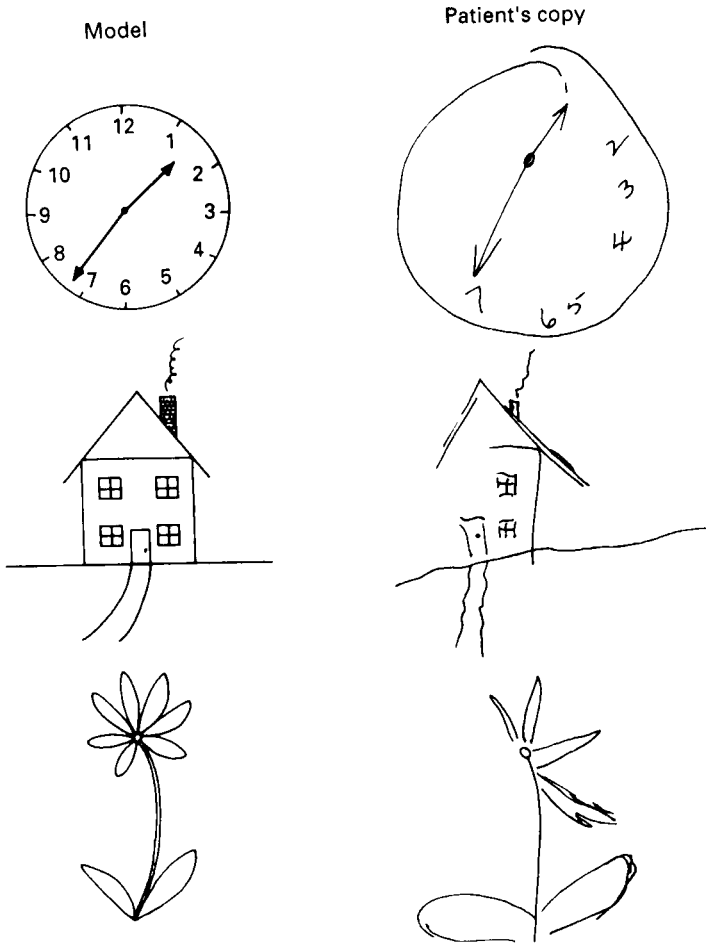

Fig. 23.5 Unilateral neglect in a patient with a lesion in the posterior part of the right hemisphere. The patient was asked to copy the drawings on the left. Reproduced with permission from Springer S P and Deutsch G (1981), *Left brain, right brain*, San Francisco, W H Freeman, 174. Copyright W H Freeman, 1981.

that they see anything there, and drawing only half a clock face (Fig. 23.5), or half a house, and in extreme cases only washing half their body, or only putting on their clothes on one side. A variant of the syndrome is UNILATERAL ASOMATAGNOSIA in which the patient cannot recognize the left half of their own body as belonging to them (asking whose leg or arm it is in the bed with them), or not recognizing that the limb is diseased (ANOSOGNOSIA). Although rare such syndromes give us valuable insights into the nature of normal processes.

The final non-dominant syndrome is CONSTRUCTIONAL APRAXIA, which is an apraxia with an inability to carry out motor actions, but unlike the dominant apraxias the deficit is not in the control of motor

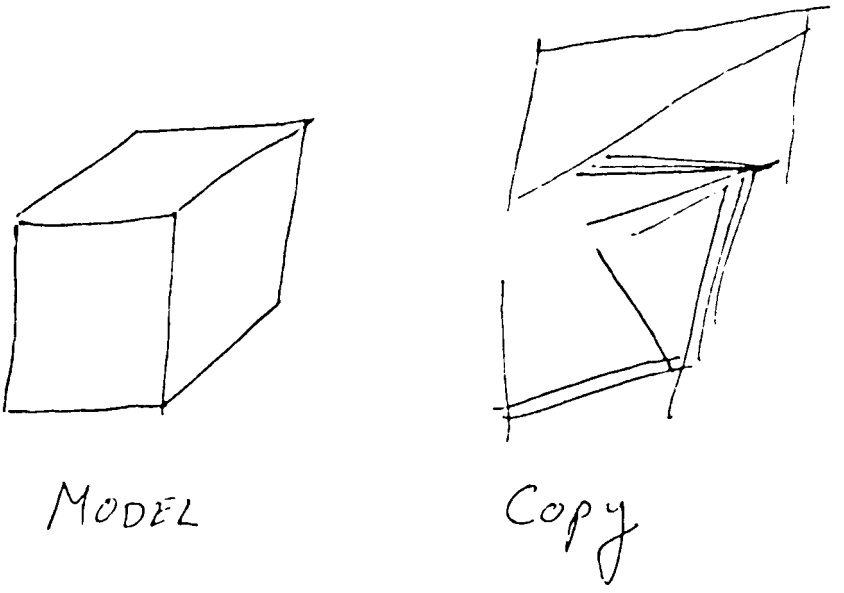

Fig. 23.6 Constructional apraxia after a lesion to the right parietal area. The patient was asked to copy the model on the left. Reproduced with permission of John Wiley and Sons, Inc, all rights reserved, from Hecaen H and Albert M L (1978), *Human neuropsychology*, New York, John Wiley.

movements but in the understanding of the three-dimensional space in which movements are made, so a patient cannot draw or copy pictures (Figure 23.6) and, in DRESSING APRAXIA, may be unable to put on clothes because of an inability to understand the apparently baffling topology of a simple garment like a cardigan. Although such defects seem mundane, it is only by observing such problems in the brain-damaged (or the very young) that we appreciate the subtleties and complexities of an everyday action that we take for granted and almost literally carry out 'without [conscious] thinking'.