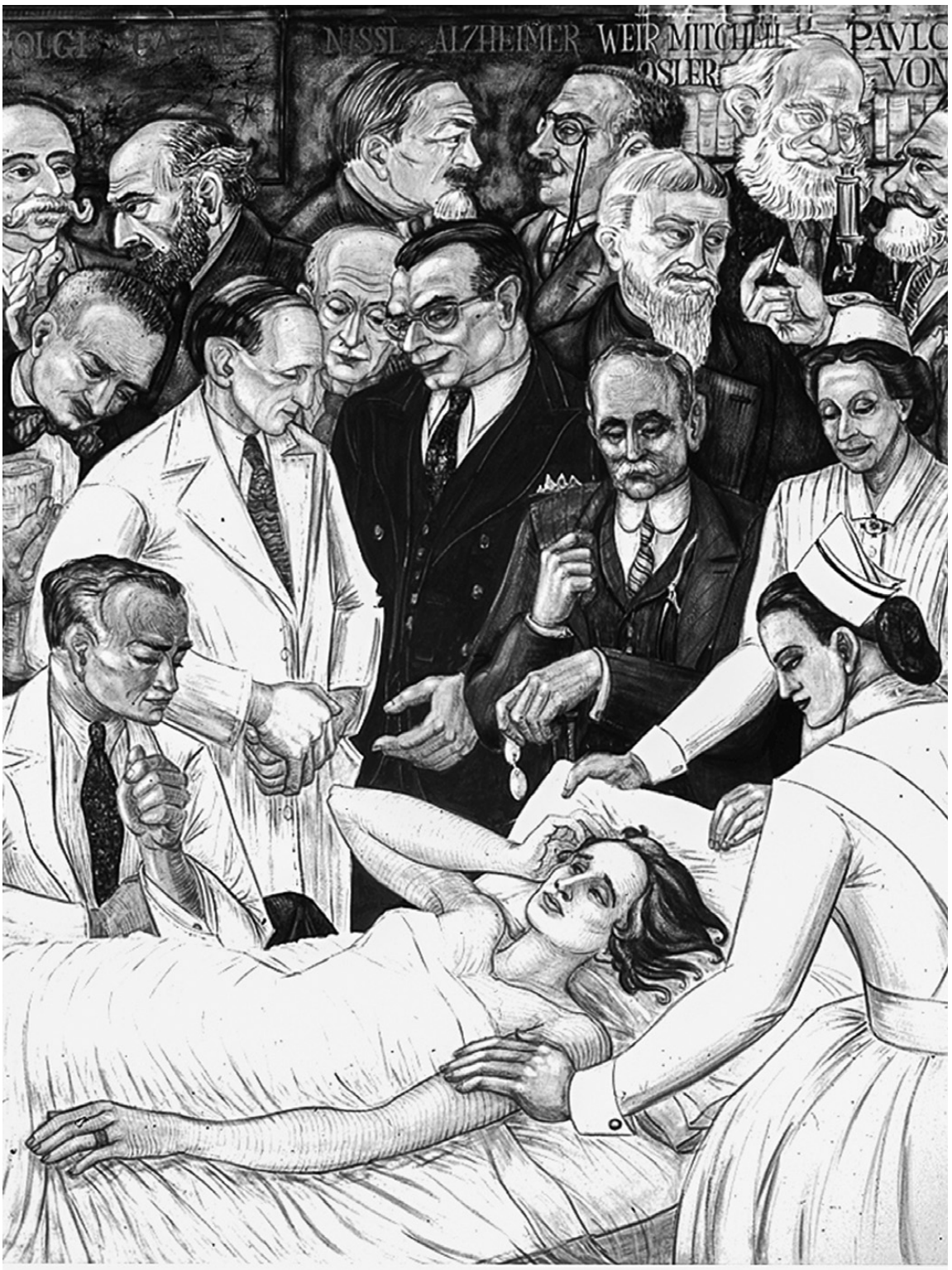

3. 'The Advance of Neurology' (1954). A detail from the painting by Mary Filer on the wall of the Montreal Neurological Institute, with Penfield and Jasper in the foreground, Mary Filer as nurse and patient, and many celebrated predecessors watching. In the back row (in this detail) are Golgi, Cajal, Alzheimer, Weir Mitchell, Osler, Pavlov and Von Monakow.

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PROLOGUE

A NOTE ON THE CONCEPT – THE IDEA – OF EPILEPSY

The concept (idea) of epilepsy – in other words, its definition, meaning and signification – needs briefly to be considered before launching into its history. I am not concerned here so much with the narrow medical definition of epilepsy, but rather with what it implied to the generality of citizens as well as to its sufferers, doctors and scientists, and how this changed over the long twentieth century. This is explored in depth throughout the next chapters and taken further at the end of this book. But it seems appropriate to offer a brief outline here.

At first sight, it may seem surprising that the concept of epilepsy would change much over time. Yet radical changes have occurred over the whole course of this period, and it is absolutely the case that the idea of epilepsy in 2020 would be *completely unrecognisable* to any citizen, doctor or patient of 1860 or 1900 or indeed later. That is not to say that any of the *physical manifestations* have altered at all; they have not, and were presumably the same in 2020 as they were in 1860 or in 400 BC when the Hippocratic collection entitled *On the Sacred Disease* was written. What has been utterly transformed in the twentieth century is how the condition is conceived, what it signifies and what it means. Any appreciation of its history, in the various periods considered in this book, must be cognisant of this fact.

As with all aspects of its history, the concept of epilepsy will vary with perspective, differing greatly, for instance, to the doctor, the scientist, the patient or the everyman. Many aspects feed into how epilepsy has been conceived, but

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at the centre are four distinct but closely interrelated issues which have played a particularly important role throughout the long twentieth century in moulding the idea of epilepsy, and which are relevant to all four perspectives:

- whether epilepsy is perceived as a *disease* (a unitary disease, or a heterogenous collection of diseases) or simply a *symptom*;
- whether epilepsy is explicable as a functional mental disorder or an organic neurological disorder;
- whether epilepsy has other inherent features apart from seizures; and
- the extent to which epilepsy is an inherited disorder.

The pendulum on each of these points has swung back and forth over the course of the century. With each reciprocating motion, the concept of epilepsy has changed, as has its meaning.

I remember lecturing some years ago to a huge gathering of epilepsy patients in India (organised by the indomitable Dr K. S. Mani). At the end of my talk, one person in the audience stood up to chastise me, enraged that I had 'referred to epilepsy as a disease'. 'It is not', I was told; 'it is a condition'. Ever since that time, with this encounter seared in my memory, this point has interested me, and by extension the question as to whether epilepsy is a disease (or condition) at all.

I do not think there is really any logical difference between a *condition*, *a disorder* and a *disease* from the scientific and medical perspective. I have yet to hear a coherent argument for differentiating the terms in these regards. But the person in the audience did have a point, for a distinction can be made on personal or societal grounds. To many individuals with epilepsy, calling it a disease implies something more serious, more biological and more fundamental. If I understood the basis of the complaint by my Indian interlocutor, it was that a condition implies that the entity is less damning and also more acceptable. The importance of terminology on framing the societal view is illustrated by this and other examples given throughout the book – not only in the use of those words now considered prejudicial but even in its their linguistic origins (for instance in the words for 'epilepsy' in some Eastern languages).

More fundamental than the semantic differentiation of disease/condition/disorder, and certainly more difficult, is the question of whether epilepsy is really a disease (or condition/disorder) at all. In other words, *does epilepsy as a disease entity actually exist?* And, if it does, what does it signify – what is its *meaning* – what is its *idea?*

THE CONCEPTS OF 'DISEASE'

One aspect of this question concerns whether there is any real distinction between epileptic seizures, the existence of which no one doubts, and epilepsy itself. In early history the question did not arise, for symptoms were considered synonymous with

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diseases (fever and skin rash, for instance). It was only after the work of Thomas Sydenham and others that a disease came to be seen as a characteristic constellation of symptoms and signs. By the nineteenth century, with the rise of the discipline of 'morbid anatomy' (pathology), the finding of pathological change had become a cardinal criterion of a disease. This criterion was supplemented after the invention of the microscope by finding histological abnormalities and then, as the twentieth century progressed, by identifying physiological, biochemical and, most recently, molecular and genetic changes. The identification of these *objective* abnormalities – loosely considered to be the *causes* of the illnesses[1] – became, in the mind of many doctors and scientists, a prerequisite for the designation *disease*. In parallel, in the last half-century molecular biology has elucidated numerous pathophysiological *mechanisms* of disease – explaining how a disease comes about. From these developments has evolved the modern *medical model* of disease in which three criteria are used to categorise an entity as a distinct disease: a unique constellation of symptoms and signs, a characteristic pathogenesis (set of mechanisms) and a characteristic causal pathology. Where no cause is found, the condition is sometimes considered 'idiopathic' (i.e. a disease unto itself, a *morbus per se*) signifying not that there is no cause but that the cause is as yet unidentified (most of these conditions are likely to have internal/congenital/inherited causes). Not all diseases fit this simple model, and boundaries can be blurred. For instance, some conditions are defined as disease because they deviate statistically from the norm (where health and disease are seen as being on a spectrum) – hypertension is one example. The greatest difficulties in applying the medical model of disease, however, relate to mental conditions (psychiatric disorders) where there is often no overt causal lesion nor clear pathogenesis, few physical signs, and where it is only subjective experience and behaviour that is considered abnormal. Accordingly, the idea that such mental phenomena are in fact diseases has been repeatedly challenged as arbitrary or even illusory (examples include the pronouncements of the anti-psychiatry movement in the 1960s, the contemporary arguments about the status of ADHD and other formulations of the DSM).

Of course, the medical model is not the only way to define disease (and again this topic is further discussed in the Epilogue of this book). Again, perspective is important. To the scientist, the condition is seen in molecular or mathematical terms, but to society the impact of epilepsy is often overwhelmingly construed in economic or legislative terms and in social and cultural attitudes. To the patients it is the fact of *being epileptic* which is often more important than just having seizures, with its effects on such aspects as social interactions, social relationships, marriage,

[1] The complexities encountered in defining 'cause' in epilepsy are discussed further on pp.606–7.

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domestic life, driving, education and employment. Their concern is with epilepsy as an illness, not a disease in the medical sense, and their actions ('illness behaviours') are often utterly unrelated to any scientific or medical concern.

THE ORIGINS OF THE MODERN MEDICAL CONCEPT OF EPILEPSY: THE WORK OF HUGHLINGS JACKSON

As we shall see, like so much else in neurology, the modern history of epilepsy can be said to have begun with the work of John Hughlings Jackson in the 1860s and 1870s. When he started out on his studies, the nature of an epileptic seizure was completely unknown. It, and epilepsy, were still considered by some to be caused by supernatural or thaumaturgic influences and both were entangled with and hardly differentiated from hysteria. Other pathogenic theories existed and the more physiologically minded physicians considered seizures to be reflexes with an origin located in the medulla or even the spinal cord. Jackson utterly dismissed these (now seemingly prehistoric) concepts. With remarkable intuition, in 1873 he proposed that the physical manifestations of an epileptic seizure were caused by *a sudden and excessive discharge* of neurons in a *localised* area of the cerebral cortex (the superficial grey matter of the brain). He saw the physical manifestations of a seizure simply to be a reflection of this disturbed physiology.[2] In Jackson's view, the seizure was a 'symptom' of this abnormal physiology. It is this singular insight that can be said to have launched the topic of epilepsy into its modern era, and since Jackson the concept of the epileptic seizure has hardly changed. At the time, there was a titanic struggle between science and religion to provide an explanatory model for the world, and Jackson's lucid explanation of epileptic seizures was a signal example of the power of science. Although clearly defining seizures, Jackson however shied away from definitively defining epilepsy – one suspects because he saw the inherent logical difficulties in doing so. To Jackson, the term epilepsy was simply shorthand for the physiochemical mechanism of seizures; in other words, the two were almost synonymous and, indeed, throughout his oeuvre he used the term *epilepsy* largely interchangeably with that of *epileptic seizure*.

THE CONCEPTION OF EPILEPSY BETWEEN THE 1880s-1930s: EPILEPSY AS A PSYCHO-HEREDITARY MENTAL DISORDER

Another conception of epilepsy arose in the late nineteenth century, with the increasingly prominent idea that heredity was the primary cause of seizures. The view grew up that only idiopathic epilepsy was the true *disease* known as epilepsy. It

[2] Jackson called this abnormal cellular 'nutrition'; today, the same concept is called 'abnormal excitation' and explained in terms of molecular biochemistry and physiology.

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was a disease that met the three criteria of a medical model of disease, with its spectrum of symptoms and signs (importantly, with symptoms extending beyond seizures), a clear-cut pathogenesis (degeneration) and a clear-cut cause (heredity). On the other hand, seizures occurring in other diseases, such as brain tumours, infections or trauma, were thenceforward considered as symptomatic seizures and not included within the rubric of epilepsy. This became the orthodox view and was held well into the 1920s.

The idea of what *Epilepsy* was had thus been transformed. It was thereafter universally considered by both medicine and the public at large to be an inherited mental disorder and a brain degeneration. The underlying inherited germ-plasm defect was known as the neuropathic trait, which was believed to cause not only seizures but also behavioural and personality failings such as criminality, amorality and sexual deviance. This was a toxic mix, and the disease engendered prejudice and hostility. In response, patients and their families sheltered in denial or concealment. Epilepsy was a condition which signalled the possession of an inherent genetic weakness, and thus was not a condition which a family would willingly admit to having. The clinical practice of the time reinforced the idea that epilepsy was a mental disorder treated, as it was usually, by psychiatrists in an asylum setting.[3] To the public there was, in effect, little distinction between epilepsy and lunacy.

In the early twentieth century, the massive public interest in heredity led to a powerful eugenic movement and the eugenicists, considering epilepsy to be an inherited organic mental degeneration, focused their ire on the personality of sufferers, their psychological defects and social worthlessness. At the same time, psychoanalysis became the predominant theory of mind, and psychoanalytical theory considered epilepsy to be inherited but of a functional nature, with both seizures and the personality defects the result of the psychological mechanism of infantile regression. These theories, both eugenical and psychoanalytical, were highly damaging. To the individual with epilepsy and to society at large in this period, epilepsy reared its head as a feared and stigmatised condition – and, furthermore, one which, because it was inherited, might overwhelm and degrade the moral fibre of Western society. In the materialistic and agnostic society of those times, the societal response was increasingly that of social exclusion and rejection. Fear that the defective germ plasm might weaken the fabric of a nation and resentment at the cost of caring for the mentally ill grew, especially in the war years. The consequences were dire; the rights of the individual with epilepsy were

[3] Neurology was in any event a young and small specialty, and its concepts converged to a great extent with those of the much larger and better-established specialty of psychiatry when it came to brain disorders. As Henry Maudsley wrote: 'Mental disorders are neither more nor less than nervous diseases in which mental symptoms predominate, and their entire separation from other nervous diseases has been a sad hindrance to progress'; Maudsley, *Body and Mind*, p. 41.

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circumscribed, resulting in widespread involuntary institutionalisation and sterilisation, and in Nazi Germany, for some, in euthanasia.

THE CONCEPTION OF EPILEPSY BETWEEN THE 1930s–1980s: EPILEPSY AS A PHYSIOLOGICAL DISORDER

The 1930s were to prove a transitional decade for the concept of epilepsy, due mainly to the growing influence of the specialty of neurology which had begun insistently to stake its claim on the condition. The divide between neurology and psychiatry widened. Neurologists (and neurosurgeons) were in general impatient with both eugenical and psychoanalytical concepts of epilepsy, and proposed that seizures were due usually to an underlying cerebral pathology, even if it could not be identified, and, in parallel, considered inheritance to be a minor part of causation. To the advanced neurologists of the period, epilepsy was not a mental disorder but a medical condition. Furthermore, they rejected the concept of inherent personality defects or behavioural features, and considered epileptic seizures to be the main manifestation of epilepsy. Opinion gathered force that *idiopathic epilepsy* was not inherited but that its cause was simply unknown, and therefore that epileptic seizures in idiopathic epilepsy were not inherently different from any other symptomatic seizures. Idiopathic epilepsy was seen simply as a variant in which the underlying pathology had not yet been detected, and not in other ways distinctive; by implication, the idea that epilepsy was a distinct disease was rejected. Given the growing number of causes identified for epilepsy, it was increasingly held that restricting the term *epilepsy* to the idiopathic condition was illogical and that it should either be replaced by a plural form (i.e. *the epilepsies*) or abandoned altogether. In the definitive textbook of neurology of its time, Kinnier Wilson deliberately entitled the chapter on epilepsy 'The Epilepsies' and, given the heterogenous causation of seizures, pondered on whether it would not be better if the term *epilepsy* was replaced by *paroxysmal disorders* or *convulsive states* to indicate 'a series of diverse conditions distinguished by the occasional occurrence of "fits"'.[4] To emphasise his point, he placed his chapter within the section titled 'Disease Conditions of Uncertain Nature'.[5] To Wilson and many neurologists of the time, a seizure was now conceived as a *symptom* of an underlying pathology, of which there were many varieties. Putting the matter into a nutshell, the senior American epilepsy specialist of the time, William Cobb, identified sixty causes of seizures, and stated in 1941: 'Because ... a great many different forms of interference with nervous

[4] Wilson, 'The epilepsies', p. 2. [5] Wilson, *Neurology*, vol. 3, p. 1469.

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integration may lead to the production of fits, and because the interfering factors are varied and even paradoxical, I believe that epilepsy cannot be called a disease ... it is a symptom of many cerebral diseases.'[6]

This was the state of play at around 1940, with the idea of epilepsy the subject of adversative opinions and unresolved debates. But then a tidal wave engulfed the seas of epilepsy and changed the concept of epilepsy again. This was the introduction of the electroencephalogram (EEG). It became suddenly possible, it seemed, to visualise the very physiological changes that Jackson had described and that occurred both during and between seizures. This radically shifted the interest to physiological theories of pathogenesis and, as Lennox famously put it, epilepsy was simply 'a cerebral dysrhythmia'.[7] Epilepsy had moved firmly into the organic neurological camp and because seizures whatever their causes seemed to have similar physiological signatures, it became a term which incorporated all conditions with seizures of any cause, not just idiopathic epilepsy.

In the aftermath of the Second World War, eugenics came to an abrupt end, discarded as an acceptable social policy. The idea that epilepsy was essentially an inherited disorder was also largely rejected (nor was there a shred of pathological evidence of an inherited lesion), as was the view that epilepsy was a mental disorder with inherent behavioural or personality features. The UN Declaration of Human Rights, coming on the heels of the Nuremburg trials, opened a new era of protection for minority groups and for disadvantaged individuals. For the first time, the voice of the patient began to be heard and lay organisations to be formed. An anti-asylum and then anti-psychiatry movement changed the public mood. In the more liberal atmosphere, societal attitudes towards epilepsy became more sympathetic. The rights of those with epilepsy began slowly to be asserted, in Western societies at least, as the condition became seen for the first time legally as well as culturally to be a medical condition like any other.

The prominence given to electrographic changes had moved the emphasis from aetiology to pathophysiology, and seizures of all causes were grouped into the single disease entity that was epilepsy. In actual fact, in this formulation (similar as it was to that of Jackson), epilepsy and seizures became again almost synonymous, albeit this time at a physiological and pathophysiological rather than clinical level. A classification of *epileptic seizures* arose in the 1960s based on both clinical and EEG appearance, and a confusingly similar *classification of the epilepsies* followed in 1989.

[6] Cited by Lennox as Cobb (1941, p. 196), although no complete reference is given (Lennox, *Epilepsy and Related Disorders*, vol. 1, p. 52).

[7] Lennox, *Epilepsy and Related Disorders*, vol. 1, pp. 51–3.

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In the 1980s, in part because of the difficulties inherent in considering epilepsy as a disease, the concept of the epileptic syndrome arose, an entity falling somewhere between the symptom and the disease. A syndrome was defined as a specific constellation of symptoms, signs and EEG features, but unlike a disease it was without a fixed aetiology (and indeed cut across aetiological categories). The concept of an epilepsy syndrome filled the void between that of the epileptic seizure and that of epilepsy.

Two further developments then shifted the medical concept of epilepsy again. First was the introduction of neuroimaging; second, the rise of molecular medicine/genetics. With these new technologies, the focus turned again onto cause. Soon, hundreds of causative conditions were recognised, not just the sixty that Cobb had identified. Neuroimaging reasserted the importance of structure as well as function, thus weakening the idea that epilepsy was a purely physiological phenomenon and leading to the view, first expressed in the 1920s, that there was often (some even claimed always) a structural anomaly or pathological lesion underlying epileptic seizures. In parallel, with the rapid development of molecular science, a whole variety of molecular and cellular physiological and chemical changes and mechanisms were identified which it was found could result in epileptic seizures. The clinical EEG came to be seen as a summation of the physiological changes and as a *diagnostic biomarker* of these rather than anything more fundamental. Furthermore, the molecular physiological and chemical processes were found to be dynamic, changing over time in an individual as epilepsy evolved into a chronic state.

By then, public attitudes had become more supportive. Discriminatory or exclusional legislation was struck out, and more resources were allocated to epilepsy. Access to medical care for those with epilepsy was provided at an unparalleled level. Perhaps for the first time in history, too, individuals were emerging from the shadows and felt able openly and frankly to write about their condition. Realistic descriptions of epilepsy, and more sympathetic attitudes, began to appear in literature and film.

THE CONCEPTION OF EPILEPSY 1990s–2020

Much changed in these decades. The 'new genetics', as it became known, first surfaced in clinical epilepsy in the mid-1990s. By 2016, more than 700 genes had been discovered in conditions which included seizures in their phenotype, and suddenly epilepsy was increasingly being conceptualised again as a genetic disorder, or at least a disorder with strong genetic influences. The old idea of the epileptic personality, which had been closely tied to the ideas of heredity, remained largely rejected, but in its place was the view that there were frequent psychiatric 'comorbidities' – actually a similar notion but with

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a different name. Epilepsy was again being conceived as a disorder with inherent and inherited behavioural and psychiatric associations in addition to seizures (and some even proposing epilepsy to be a 'spectrum disorder'). In these ways, the 'new' ideas about the inheritance of epilepsy were a modern reworking of the concept of the neuropathic trait. Furthermore, neurology and psychiatry grew closer together (united under the banner of neuroscience). And although epilepsy was still very much considered a neurological rather than a mental disorder, this distinction was becoming increasingly blurred.

And so by 2020 there were a number of conceptual parallels with epilepsy in the late nineteenth/early twentieth century. Of course, much also was different. The rights of the person with epilepsy were not infringed in 2020 as they were in the early twentieth century, and societal attitudes were not remotely as hostile (at least in Western societies). Stigma remained a problem in 2020, but of orders of magnitude less than in the pre–World War II years, and people with epilepsy were able to be far more open and less fearful about their condition. Nevertheless, the parallels should not be ignored. Without vigilance, one can see how easily eugenic concepts might creep again into epilepsy and how public attitudes may again harden. And although by 2020 the meaning of epilepsy was different from that at the beginning of this story, there is a clear sense of circularity; concepts have been recycled and ideas have re-emerged.

SHOULD EPILEPSY STILL EXIST?

A final thought, taken up again at the end of this book, is that, from the scientific and medical perspectives, because of the multiplicity of aetiologies and molecular abnormalities and mechanisms, and the great variation in the clinical symptomatology and context, the idea that there is a clear-cut disease called epilepsy has become increasingly untenable. Epileptic seizures have come to be perceived as a manifestation of myriad processes and causes, and no one doubts their existence. But, from the medical perspective, the only way in which a disease called epilepsy could be considered valid is to define it as a 'state' in which there was a 'propensity to have recurrent seizures' – a wholly circular and unsatisfactory way of conceptualising a disease, and one which meets none of the aforementioned criteria of the medical model.

It is from the societal and personal perspectives, however, that the use of the term epilepsy has more legitimacy, as a useful shorthand to signify those who suffer from seizures – regardless of cause or pathophysiology. The downside, of course, with this use of the term is that it comes with much historical baggage, prejudice and stigma.

Given that there is little justification for considering epilepsy to be a disease on medical grounds, might it not be better to dispose of the

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term *epilepsy* altogether; to refer to individuals simply as suffering from epileptic seizures and not as persons with epilepsy (or, worse, 'epileptics'). These are issues debated in the final paragraphs of this book.

It is with these thoughts in mind, framed by the contextual background of the shifting conceptions of epilepsy, that we approach the narrative history of epilepsy in the long twentieth century, the topic of the next section.

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