1. Introduction

1.1 Creutzfeldt-Jakob Disease (CJD) is a rapidly progressive and ultimately fatal disorder of the central nervous system belonging to a group of disorders known as transmissible spongiform encephalopathies (TSEs). As described in vol. 2: Science, CJD can be divided into three categories: sporadic, iatrogenic and inherited. Sporadic CJD (ie, occurring in scattered instances) occurs in all countries, with a random case distribution and an annual incidence of one per million. Iatrogenic CJD arises from accidental exposure to the CJD agent through medical or surgical procedures. Inherited CJD, or familial CJD, accounts for 15 per cent of all CJD cases and is now known to be associated with prion protein gene mutations in the germ line. No such mutations in the germ line are present in sporadic or iatrogenic CJD, although somatic cell mutation of the prion protein gene is thought to be a possible cause of sporadic CJD.

1.2 In this volume we chart the development of CJD surveillance in the UK leading up to the identification and announcement of a ‘new variant’ form of CJD in March 1996. Chapter 2 presents a history of CJD surveillance and the understanding of the epidemiology of CJD up to 1990, when the CJD Surveillance Unit (CJDSU) was established in Edinburgh. This describes the early work by Professor Bryan Matthews and Dr Robert Will in Oxford, from 1975, and the decision to reinstitute the surveillance of CJD following the emergence of bovine spongiform encephalopathy (BSE) in 1986.

1.3 The CJD surveillance programme was established in May 1990, under the direction of Dr Will at the CJDSU, Western General Hospital, Edinburgh. The initial aim of this programme was to identify any change in the pattern of CJD that might be attributable to the emergence of BSE. Chapter 3 describes the establishment of the Unit, its role and the resources available.

1.4 Chapter 4 covers the studies conducted by the CJDSU, the development of the questionnaire used to obtain clinical and epidemiological information, and the results published in the Unit’s annual reports.

1.5 Chapter 5 contains a chronological account of the events preceding the official announcement on 20 March 1996 regarding the possible link between BSE and a new variant of CJD. This chapter describes the reports of several suspect cases of CJD in farmers and young people that caused concern amongst government officials dealing with the BSE issue, and eventually led to the official announcement. The last section of this chapter includes the final outcome of each of the suspect cases in farmers and young people, and gives a brief summary of the main findings of the CJDSU from 1996 onwards.

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1 Prion protein – an abnormal form of the prion protein is present in transmissible spongiform encephalopathies such as CJD. The prion protein gene and its associated protein product are normal constituents of the cell. In a poorly understood mechanism, normal cellular prion protein undergoes a transformation to a disease-inducing form. Mutations of the prion protein gene are so far the only established originating cause for the conversion of the normal molecule into the disease-producing molecules

2 Germ line – a group of cells in animals that give rise to the reproductive cells. The genome of the animal, as contained in these cells, along with any mutations in them, can be passed on to offspring

3 Somatic cell – any cell that is not involved in reproduction, ie, not a germ line cell
Chapter 6 examines the clinical features of vCJD and details its clinical diagnosis by the medical profession. This chapter also discusses the treatment and care of vCJD, addressing some of the issues that were and are still being faced by victims of the disease and their families, both during illness and following death. It contains information contributed by family members of the victims as well as from those providing care. Material from reports produced on the subject is also included. Issues covered include the provision of information, the standard of care available in hospitals and assistance given for caring at home, difficulties surrounding post-mortem and burial, and the difficulties concerning death certification and inquests. The chapter concludes with a discussion on the diagnosis, treatment and care of victims of the new disease.