The Proceedings of the 21st Annual History of Medicine Days Conference 2012

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The University of Calgary Faculty of Medicine, Alberta, Canada

Edited by

Kelsey Lucyk, Aleksandra Loewenau and Frank W. Stahnisch

Cambridge Scholars Publishing

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This book first published 2016

Cambridge Scholars Publishing

Lady Stephenson Library, Newcastle upon Tyne, NE6 2PA, UK

British Library Cataloguing in Publication Data
A catalogue record for this book is available from the British Library

ISBN (10): 1-4438-9765-5
ISBN (13): 978-1-4438-9765-5

The publication of the 21st Annual History of Medicine Days Proceedings was supported by a 2012 annual conference grant from AMS (Associated Medical Services, Inc.). The content is solely the responsibility of the authors and does not necessarily represent the official views of Associated Medical Services, Inc.

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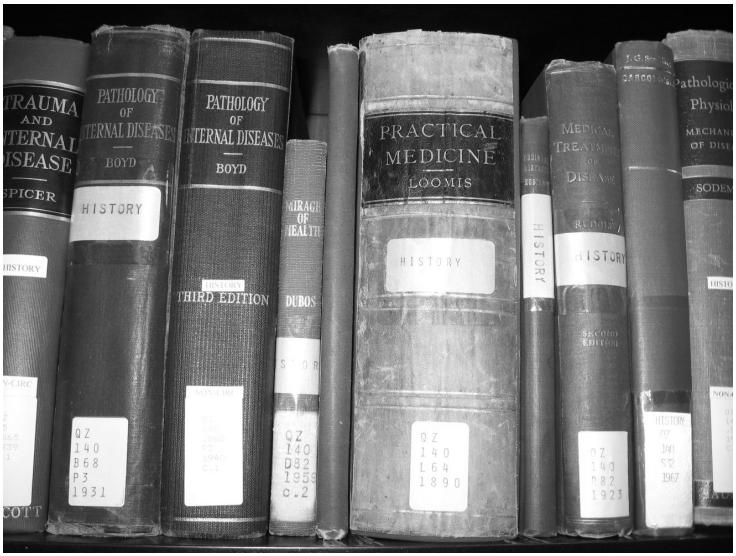
PREFACE AND ACKNOWLEDGEMENTS

The time for the preparation and editing of this twenty-first volume of the University of Calgary History of Medicine Days Conference Proceedings has become a source of great joy and satisfaction for the editorial team throughout.[1] The 21st History of Medicine Days (HMDs) conference took place on Friday March 9, 2012 and Saturday March 10, 2012 in the Libin Theatre of the University of Calgary's Faculty of Medicine in Alberta, Canada. Here, undergraduate and early graduate students from across Canada, the United States, United Kingdom and Europe gave paper and poster presentations on a wide variety of topics from the history of medicine and health care. In this preface, the editors would like to express their particular and sincere thanks again to all of the presenters and event participants of the 21st HMDs student conference in 2012. The event was graciously introduced by the Dean of the University of Calgary's Faculty of Medicine, Dr. Thomas E. Feasby, and the Dean of the newly amalgamated Faculty of Arts, Dr. Kevin McQuillan. Both gave their warm welcoming addresses to all delegates and participating students from universities and medical at the University of Calgary.

The resulting chapter contributions in the first part of this volume provide many historical insights, which could be gained from the 2012 conference event in Calgary. They are assembled here for the readers of the current HMDs Proceedings volume. As for this year's HMDs conference, the selected papers, which are included in the current volume, particularly comprise contributions on Historical Perspectives on Social Determinants of Health; Perspectives on Nursing History; and Medical Education. Further content areas include Epilepsy, Hysteria and Laughter; History of Neuroscience; History of Clinical Research; as well as History of Neuro- and Psychosurgery; Interdisciplinary perspectives on the history of medicine close this volume, such as Medical Philosophy and Break-Throughs and Disasters. All in all thirty-nine oral presentations and ten

[1] The new specialized series of the History of Medicine Days conference with Cambridge Scholars Publishing began in the year 2012. See also Lisa Petermann, Kerry Sheng–Sun and Frank W. Stahnisch (eds.), *Proceedings of the 18th History of Medicine Days Conferences at the University of Calgary, AB* (Newcastle upon Tyne: Cambridge Scholars Publishing, 2012).

posters were presented at the 2012 conference. Furthermore, both an exhibit of objects from the Alberta Health Services (AHS) historical archive collection (Chief Curator: Mr. Dennis Slater) and from the Mackie Family Collection in the History of the Neurosciences at the Health Sciences Library of the University of Calgary Faculty of Medicine (Library Director: Mrs. Susan Powelson) were made available for viewing at the entrance to the Libin Theatre in the Health Sciences Centre of the Faculty of Medicine in Calgary.



1-1 Pathology books from the Mackie Family Collection in the History of Neuroscience that were exhibited at the 21st History of Medicine Days Conference in March 2012. Photograph taken in the History of Medicine Room by Frank W. Stahnisch prior to the conference, 2012. Source: Courtesy of the Health Sciences Library of the University of Calgary, Alberta, Canada.

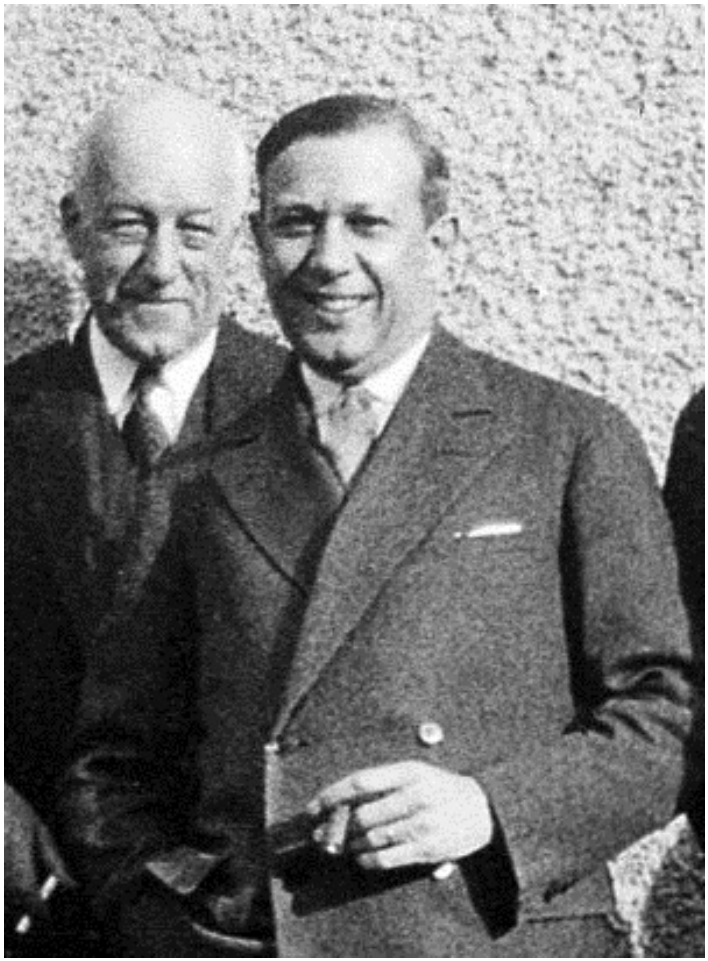
Altogether student from the University of Calgary delivered one fourth of the presentations at the 21st History of Medicine Days conference. Many local students also actively volunteered on various organizing committees for the conference as well, which included hosting and billeting external student presenters in Calgary, as well as aiding with the organization of the program, and many other preparatory roles. Without their most valuable and gracious help, the organization of the conference

would not have been as smooth and successful as it was. About one hundred and fifty students and faculty colleagues attended the individual sessions from the University of Calgary campuses. Additionally, many faculty colleagues from both the Faculty of Medicine and the Faculty of Arts supported the HMDs by reviewing conference abstracts that were submitted.

They also contributed significantly to the chairing and judging of the numerous paper and poster presentations that were delivered at the conference. As well, faculty colleagues were also involved in the organization of a specialized, interactive session entitled, "What to do with (Hi-)stories? – Book Review Presentations from Students in the Calgary Narrative Medicine Program." This special session was co-organized by faculty colleagues from the Department of Paediatrics and the Narrative Medicine Interest Group at the Faculty of Medicine, and was chaired by Professor Ian Mitchell.

The conference's keynote lecture, entitled "Henry Sigerist's Advocacy of Social Medicine and Universal Health Insurance in North America, 1932–1947," was given by the renowned historian of public health, Professor Theodore M. Brown from Rochester University in New York, who presented a broadly received and well-appreciated address to the conference. Professor Brown has kindly rendered his talk into a chapter contribution for this 21st History of Medicine Days Proceedings volume. He has also made important archival materials available for this publication, which build on his important earlier work on the leading Swiss-American émigré historian Henry E. Sigerist (1891–1957) at Johns Hopkins University in the United States.[2] In 2012, Dr. Brown's invited keynote lecture was again delivered as a co-sponsored event by the O'Brien Institute for Public Health (OIPH), the Calgary History of Medicine Society (CHOMS) and the Science, Technology, Environment and Medicine Studies (STEMS) colloquium, for which the organizers of this event were all very grateful.

[2] Theodore M. Brown, "Friendship and Philanthropy: Henry Sigerist, Alan Gregg and the Rockefeller Foundation," in: Elizabeth Fee and Theodore M. Brown (eds.): *Making Medical History. The Life and Time of Henry E. Sigerist*, 288–311 (Baltimore: Johns Hopkins University Press, 1997).



1-2 Medical historian Henry E. Sigerist (1891 – 1957), pictured April 27, 1929 with Swiss medical historian, Arnold C. Klebs (1870 – 1943).
This file (PHO No 15183) comes from Wellcome Images, a website operated by Wellcome Trust, a global charitable foundation based in the United Kingdom.

Henry E. Sigerist, a former Johns Hopkins University's social historian of medicine, is often acclaimed as a founding figure of comparative health systems analysis, based on the influential books he wrote on the newly developing public health programs in the Soviet Union from 1917

onwards,[3] and also his writings on the status and scope of medicine and health care in the United States.[4] Professor Brown's chapter in this volume delves particularly into the health systems investigations and public health research activities of Sigerist and sheds further light on how his work contributed to the discussions that were emerging in Canada to create a generally accessible public health care system. In Canada, publicly funded health care originated primarily in Alberta's neighbouring province of Saskatchewan, and is a development that has long received the attention of scholars in the field of the history of medicine and public health. Placed in its political and social context, it is fitting – as Dr. Brown has pointed out – that Sigerist was invited to become the Chair of a local provincial committee tasked with researching the status and scope of the health care services in the province of Saskatchewan, which had existed since 1933. Tommy Douglas (1904—1986), the new leader of the union-based Cooperative Commonwealth Federation (CCF; which later became the New Democratic Party in 1961) who spearheaded new health care initiatives in Western Canada, led this initiative.

This survey of health care services in Saskatchewan preceded the institutionalization of a taxation-based health care system that was implemented in the province and promoted to the federal Government of Canada in September 1944. The program, for which Sigerist had worked and advocated for, foresaw the development of free hospitalizations for all inhabitants of the province should further lead to an augmented and comprehensive health care system. Sigerist, a Swiss born and German-trained historian of medicine, became involved in the Saskatchewan-based committee due to his increasing renown and influence in American and Canadian discussions about the future of the existing health care systems. During the 1930s and 1940s, former Saskatchewan Premier, Tommy Douglas, had read many of Sigerist's books and also attended to the advice of his public health-oriented physicians relating to his new provincial government. For them, Sigerist was the international leader in the field of comparative health systems research and health policy counselling. Eventually, Sigerist came to the Canadian prairies to examine the local hospital and health care institutions' plans for future health care reform in Western Canada and later, potentially, in the whole country.[5]

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[3] Henry E. Sigerist, *Socialized Medicine in the Soviet Union* (New York: W. W. Norton & Co., Inc., 1937).

[4] Henry E. Sigerist, *American Medicine* (New York: Norton, 1934).

[5] Jacalyn Duffin, "The Guru and the Godfather: Henry Sigerist, Hugh MacLean, and the Politics of Health Care Reform in 1940s Canada," *Canadian Bulletin of Medical History* 9 (1992), pp. 191-218.

Sadly, the 21st History of Medicine Days conference in Calgary was overshadowed by the severe illness that had struck Dr. B. William Shragge, which the organizers had been informed shortly before the conference itself. Dr. Shragge, whom was referred to by his friends and colleagues simply as "Bill," had been the dedicated CEO of Associated Medical Services (formerly known as the Hannah Foundation for the History of Medicine) and a fervent supporter of the Calgary History of Medicine Days. For many years, Bill regularly attended and greatly enjoyed this event. It came as a shock to the organizers of the conference when Bill sent his regrets this year, as he was bed-struck by the time of notice. Unfortunately, the bad news turned worse and Bill eventually succumbed to his enduring illness and passed away shortly after the conference on April 20, 2012.[6] At the age of 64, Bill has left us much too early. He has left behind his contributions to and is also mourned by the Canadian medical community, the Canadian communities in history of medicine and the medical humanities, but above all, he has left his dear family: his beloved wife Phyllis, his greatly treasured children, Marla, Andrea, Avi, Benjamin, and Rebecca, and his first grandson, Max, who was born in 2010. There was rarely a social gathering with Bill in attendance where he did not mention the families of his friends and colleagues or imparted stories about his own. The History of Medicine Days in Calgary were no exception from that rule. Bill took great interest in the organizers' activities, their work lives and families, and was often seen chatting together with the students and presenters until late, to hear about their studies, career plans, and situation. Whatever the pressures and constraints of his working life, it was clear to all who knew him that Bill's family life was most precious to him. He was "a whirlwind" in his professional life as a physician and medical administrator. The Calgary HMDs conferences are greatly indebted to him and his enthusiasm and will miss him dearly.

Bill was born in Winnipeg, Manitoba, which he jokingly referred to as "Winterpeg." Likewise, it was minus 30°C on the day before the HMDs conference in the preceding year. He often referred to "his refreshing Canadian winters" and how he greatly missed them when working south of the border. Bill had received his medical education in cardiovascular and thoracic surgery, as well as in critical care medicine from the University of Manitoba in 1976. That same year he also passed the exams

[6] An official obituary can be viewed at: Niagra Health Services: "Former NHS Chief of Medical Staff Dr. Bill Shragge Dead at 64," *Newsbulletin Nigara* (April 21, 2012) (http://www. bulletnewsniagara.ca/2012/04/21/former-nhs-chief-of-medical-staff-dr-bill-shragge-dead-at-64/); <accessed May 20, 2012>

for a Royal College Fellowship. Subsequently, he moved to Birmingham, Alabama, for a postgraduate fellowship of two years, which he described as "the most demanding climate I ever worked in." In 1978, he became a faculty member of McMaster's Department of Surgery where he also was Chair between 1985 and 1990. Complementary to his career in academic surgery, Bill developed a strong interest in medical education that became a personal vocation for him: he contributed to the McMaster Undergraduate Medical Education program, supported student activities through Associated Medical Services, was involved in the Canada-wide History of Medicine Days conferences in Calgary,[7] and was most recently engaged in the medical humanities through the AMS Phoenix program entitled, "A Call to Caring." [8] All of this dedication spoke to his enthusiasm for medicine and health care. As he told the author (FWS) in March of last year, "The future of medical care depends on building an inter-professional community."[9]

Throughout his career, Bill held numerous leadership positions. Among others, he was the founding Chief of Staff of the Hamilton Health Sciences Corporation (1996), Chief of Staff of the Niagara Health System (2001), and Chief of Staff and Vice-President for Academic Medical Affairs of St. Peter's Hospital in Hamilton (2004). In July 2009, Bill became the new CEO of Associated Medical Services, a position in which he endeavoured to sustain the organization's alignment with a number of history of medicine groups and activities in Canada. Included in these connections were the Canadian Society for the History of Medicine, the History of Medicine Days conferences in Calgary, and the grant programs for students through AMS. Bill further accentuated AMS's support for a broader medical humanities initiative, one in which he perceived that the history of medicine would play a very significant part. It was astounding to know that even though he had been diagnosed with lung cancer in December of 2010, he continued to say, "we're just at the beginning of

[7] University of Calgary, "20th Anniversary Panel of the Calgary History of Medicine Days," in University of Calgary Faculty of Medicine (ed.): History of Medicine Days Conference Program (2011): 1 (http://www.hom.ucalgary.ca/system/files/History+of+Medicine+Days+2011+-+PROGRAM+-+February+25,+2011-1.pdf); <accessed May 22, 2012>.

[8] Associated Medical Services, *The AMS Health Professional Initiative* (Synthesis Document), Toronto, ON: Associated Medical Services, 2011.

[9] B. William Shragge, "The Genesis and Development of AMS's Projects: A Call to Caring," in: Aleksandra Loewenau, Kelsey Lucyk, and Frank W. Stahnisch (eds.): *Proceedings of the 21st History of Medicine Days Conferences at the University of Calgary, AB*, 45–52 (Newcastle upon Tyne: Cambridge Scholars Publishing, 2012).

this project!"

Asymptomatic for one year, he worked long days and travelled to many meetings throughout North America. However, his health deteriorated early in 2012 and in April he became hospitalized, leaving us all a few weeks later. Bill's passing leaves an enormous void in the Canadian history of medicine community as well as in the broader community of medical education. Those who had a chance to meet and talk with him appreciated the warmth of his character and the great personal interest he took in everything and everyone connected to medical education. The openhearted manner, in which he conversed, particularly with students, encouraged them to uphold their ideals and follow their dreams. We feel the heavy loss of a fine colleague and good friend.[10]

Looking at the current HMDs Proceedings Volume, the editorial team is very grateful that a solid number of ten manuscript contributions could be included in this edited collection.[11] Among these contributions is also an interesting communication in the history of medicine, authored by the Chief Curator of the Alberta Health Sciences' Historical Archive Collection, Mr. Dennis Slater. In this contribution, he describes how those interested in artifacts and objects from the history of medicine and science in Western Canada can substantially benefit from the resources and collections of the Alberta Health Services Archives & Historic Collection in Calgary.[12] Moreover, this volume includes the abstracts of all 2012 conference presentations in a separate appendix. Furthermore, this volume has been illustrated with images and diagrams pertaining to the various topics from the history of medicine as they are assembled here. Throughout the 21st conference of the History of Medicine Days in 2012 at the University of Calgary, research enthusiasm, oratory, and audiovisual competence of the speakers were again of a very high quality. The conference audience was not only greatly entertained, but the local and national delegates contributed further to a highly stimulating and engaging discussion throughout the whole event. The proceedings editors are

[10] Frank W. Stahnisch, "obituaries / annonces nécrologiques – Dr. B. William Shragge," *Canadian Bulletin of Medical History* 29 (2012), pp. 43-45.

[11] All of the contributions in these proceedings, for which the volume editors have received explicit *Copyright Transfer Forms* and *Author Consent Forms*, will also be made available online through the University of Calgary Internet Repository *Prism*: History of Medicine Days Community Homepage: http://dspace. ucalgary.ca/handle/1880/47439.

[12] Alberta Health Services, "Alberta Health Services Archives & Historic Collections" (http://www.albertahealthservices.ca/info/Page4306.aspx); <Accessed on September 15, 2016>.

grateful to all participants for their active contributions and support, which helped to make this academic conference a great success. We had greatly commiserated that AMS Chief Executive Officer, Bill Shragge, had not been able to attend our conference, due to his diminishing health. We were even more saddened to hear later, when the editing work on this conference volume had started, that he had succumbed to his prolonged illness and passed away shortly thereafter. Both the publication of the Proceedings Volumes and the organization of the 21st History of Medicine Days at the University of Calgary would not have been possible without the lasting and substantial financial support through Associated Medical Services in Toronto along with the Alberta Medical Foundation in Edmonton, for which we continue to be very grateful. This publication was supported by a 2012 program grant from AMS (Associated Medical Services, Inc.). The content is solely the responsibility of the authors and does not necessarily represent the official views of Associated Medical Services, Inc.

We would also like to thank the O'Brien Institute for Public Health in Calgary and the Faculty of Medicine at the University of Calgary for their continued and important support. We likewise extend our warm thanks to Beth Cusitar for the important additional editorial help she provided in finalizing this manuscript, as well as her tireless efforts in organizing the conference and contributing greatly to its success for yet another year. In addition, we express our gratitude to the staff at Cambridge Scholars Publishing, who have been instrumental in the *Proceedings of the History of Medicine Days Conference* Volumes as a full History Series with their press in Newcastle upon Tyne.

Frank W. Stahnisch,
Kelsey Lucyk,
Aleksandra Loewenau,

(University of Calgary, January 7, 2016)

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HISTORICAL PERSPECTIVES ON THE SOCIAL DETERMINANTS OF HEALTH

WHILE ENGLAND ATE: GOVERNMENT AND THE RISK OF BSE, 1979 – 1996

MICHEL SHAMY

SUMMARY: In 1984, Britain's cattle began to fall victim to a new disease, one that would eventually be recognized as Bovine Spongiform Encephalopathy (BSE), or "Mad Cow" Disease. It was not until March of 1996 that the government of Britain officially recognized BSE as a threat to human health in the form of a new syndrome called Variant Creutzfeldt-Jakob Disease (vCJD). How did the government quantify and manage the potential impact of BSE on human health prior to 1996? I will argue that the British government's policies implicitly accepted that BSE could be spread to humans; simultaneously, official pronouncements denied that any risk existed. The uncertainty produced by facing a new disease allowed biases and assumptions to delay and undermine the efficacy of the government's own policies. By comparing Britain's response to BSE to that of the Canadian government, as well as to Britain's management of cattle plagues in the Nineteenth Century, we recall familiar tensions between public safety and financial interests, between the communication of information and the avoidance of panic, and surrounding the distribution of authority among experts, bureaucrats, and politicians.

KEYWORDS: Bovine Spongiform Encephalopathy, variant Creutzfeldt-Jakob Disease, Great Britain, Risk, Uncertainty, Public Health, Canada

PRECEPTOR: Dr. Frank W. Stahnisch
INSTITUTION: University of California, San Francisco

Introduction

In the mid 1980s, British cattle began to succumb to a new and frightening disease. Officially designated Bovine Spongiform Encephalopathy (BSE), this condition became famously known worldwide as "Mad Cow Disease." Very soon after its initial description, medical professionals and government officials began to consider the risk BSE might pose to humans. Within a decade, young people across Britain began dying from a unique syndrome which, it was thought, might be related to Mad Cow Disease. By the end of the 1990s, extensive medical evidence proved that this new human disease, variant Creutzfeldt-Jakob Disease (vCJD), was in fact acquired by consuming beef infected with BSE.

This chapter intends to provide an historical analysis of the means by which the British government considered and responded to the risk posed by BSE to humans in the years 1979 – 1996. I will argue that the measures enacted by the government implicitly accepted that BSE could be spread to humans, while ministers and officials repeatedly denied that any risk existed. In light of the uncertainty surrounding the science of BSE, the biases and assumptions of politicians and bureaucrats delayed and undermined the government's own policies.[1]

The case of BSE and vCJD in Britain recalls familiar themes from the history of medicine. In Britain's management of the BSE risk we can appreciate the tensions that existed between (1) public safety and financial interests; (2) the desire to communicate information and avoid panic; and interests surrounding (3) the distribution of authority among experts, bureaucrats, and politicians. Despite medical uncertainty, it was still necessary to make decisions to manage BSE. Where medical uncertainty exists, it is likely that a government's own biases and assumptions will direct policy decision, although these influences are rarely recognized.

This discussion is organized into five sections. First, I will briefly review the scientific and medical aspects of BSE and its related diseases. Second, I will consider the rise of BSE and the government's early assessment of BSE's risk to humans. Third, I will assess the 1989 report of the Southwood Working Party and the consequences of its recommendations. Fourth, I will analyze the government's response to the development of new variant Creutzfeldt-Jakob disease in humans. Fifth, I will use comparative perspectives to illuminate the British government's BSE policies.

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[1] For the issue of historical responses to public health issues by politicians and bureaucrats, see for example: George Rosen, *A History of Public Health* (Baltimore: Johns Hopkins University Press, 2015): 350–359.

Despite an extensive body of literature on BSE, there is little written on BSE and vCJD within the broader history of medicine. Most authors who have written about BSE in Britain have focused on the disease's political, economic, or biomedical aspects; by adopting a medical historical approach, these various perspectives can be unified.[2] Despite having summarized valuable and wide-ranging testimony, the four-thousand page *Phillips Inquiry*, published in 2000, remains a biased secondary source influenced by the political and economic environment within which it was written. Since its publication, the report's authors have frequently been criticized for their unwillingness to apportion blame for the BSE crisis to the scientific experts and government officials directly implicated.[3] Many BSE experts (for example Paul Brown, John Collinge, and James Ironside) have written eloquently about the origins of the disease, but their narratives are necessarily coloured by their association with its management.[4,5,6] In the recent book *Rethinking the BSE Crisis: A Study of Scientific Reasoning under Uncertainty*, Nottingham Trent University Professor of Linguistics, Louise Cummings, provides an epistemic analysis of the argumentation and the process of decision-making involved in the management of BSE and variant CJD, though her argument is limited by its reliance on the text of the *Phillips Inquiry*.[7] Therefore, the need remains for an historical analysis of the medical decision-making involved in the management of BSE between 1979 and 1996 and this chapter will attempt to serve that purpose. As a practicing neurologist who has cared for patients with Creutzfeldt-Jakob Disease, I admit that I may be biased by my own experience with the suffering I have witnessed at the hands of the spongiform encephalopathy. Yet I disclose that it is with this perspective that I approach my research.

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[2] John Fisher, "Cattle Plagues Past and Present: The Mystery of Mad Cow Disease," *Journal of Contemporary History* 33 (1998): 215–228.

[3] Jonathon Carr-Brown and Senay Boztas, "A Culture of Secrecy that Risked our Lives," *Times of London*, October 29, 2000, Features, 5.

[4] Paul Brown, "Mad-cow Disease in Cattle and Human Beings: Bovine Spongiform Encephalopathy Provides a Case Study in How to Manage Risks While Still Learning the Facts," *American Scientist* 92 (2004): 334–341.

[5] John Collinge, "Human Prion Diseases and Bovine Spongiform Encephalopathy," *Human Molecular Genetics* 6 (1997): 1699–1705.

[6] Graham Mackay, Richard Knight, and James Ironside, "The Molecular Epidemiology of Variant CJD," *International Journal of Molecular Genetics* 2 (2011): 217–227.

[7] Louise Cummings, *Rethinking the BSE Crisis: A Study of Scientific Reasoning under Uncertainty* (London: Springer, 2010).

2-1 Plaque commemorating BSE victims in London, England.
Copyright Matt Brown
Source: https://www.flickr.com/photos/londonmatt/16448174166

The Transmissible Spongiform Encephalopathies

Bovine spongiform encephalopathy is one member of a family of diseases known as the Transmissible Spongiform Encephalopathies (TSE), which afflict humans and a number of mammalian species including cows, sheep, deer, and elk.[8] The TSEs are rapidly progressive brain diseases that invariably lead to death. Pathologically, they share the feature of spongiform changes in the brain, resulting in a microscopic "Swiss cheese-like" appearance. The oldest known TSE is "scrapie," a disease of sheep first identified in Scotland in 1732. Because scrapie had never been shown to infect humans, it was assumed that BSE would behave in a similar way.[9]

Although TSEs have been known for over two hundred years, the

[8] John Collinge and Stanley Prusiner, "Terminology of Prion Diseases" in *Prion Diseases of Humans and Animals*, Stanley Prusiner et al. (Chichester: Ellis Horwood Ltd., 1992).
[9] Raymond Bradley, "Bovine Spongiform Encephalopathy: The Current Situation and Research," *European Journal of Epidemiology* 7 (1991): 532–544.

nature of their transmissibility was only recently explained. For much of the twentieth century, it was assumed that a "slow virus" was the causative agent behind the TSEs. Beginning in the 1980s, American neuroscientist Stanley Prusiner (b. 1942) began to argue that TSEs were caused by an infectious protein, *the prion*. Prions (designated PrP^{c}) are naturally occurring proteins found in the central nervous system. Under circumstances that remain unexplained, prions can obtain a misfolded shape (PrP^{sc}) that is, in and of itself, transmissible to other prion proteins. This abnormal prion is highly contagious and is extremely resistant to degradation by usual sterilization techniques.[10] Dr. Prusiner's theory was ultimately proven correct and he was awarded the 1997 Nobel Prize for Physiology or Medicine at the height of the BSE epidemic.

The incidence of TSEs in humans is stable across most studied populations, at about 1 per million per year, of which the vast majority are caused by Creutzfeld-Jakob Disease. Creutzfeldt-Jakob Disease was first described in 1920, and presents as a rapidly-progressing dementia. Mean survival is four to eight months; ninety percent of patients are dead within one year. The peak age of onset is fifty-five to seventy-five years. The diagnosis is firmly established through autopsy, although characteristic changes on magnetic resonance imaging (MRI) scan and electroencephalography (EEG) are highly sensitive and specific.[11] Other human TSEs include Fatal Familial Insomnia (FFI), Gerstmann-Straeussler-Scheinker (GSS) syndrome, and Kuru. Notably, Kuru was disseminated among the *Foré* tribe of Papua New Guinea, where it was associated with the ceremonial consumption of the brains of dead relatives.

Beginning in 1993, a new clinical syndrome reminiscent of Creutzfeldt-Jakob disease was identified among young people in Britain.[12] As of 2011, there have been two-hundred and eighteen cases of variant Creutzfeldt-Jakob Disease (vCJD), mostly in the United Kingdom (UK) and France, and some in Canada and the United States.[13] The disease is

[10] David Ellison, Seth Love, Leila Maria Cardao Chimelli, Brian Harding, James Lowe, Harry Vinters Sebastian Bradner, and William Yong, *Neuropathology: A Reference Text of CNS Pathology*, 2nd Edition (Edinburgh: Mosby, 2004), 585.
[11] Michael Geschwind, "Rapidly Progressive Dementia: Prion Diseases and Other Rapid Dementias," *Continuum: Lifelong Learning in Neurology* 16 (2010): 31–56.
[12] Robert Will, James Ironside, Martin Zeidler, Simon Couseus, Kavitha Estibeiro, Annick Alpévovitch, Stanley Poser, Maurizio Pocchiari, Albert Hofman, and Peter Smith, "A New Variant of Creutzfeldt-Jakob Disease in the UK," *Lancet* 347 (1996): 921–925.
[13] Mackay, "Molecular Epidemiology," 218.