1.1	ld number	
1.2 ⁻	country Belgium (1); France (2); Germany (3); Italy (4); the Netherlands (5); United Kingdom (6); Spain (7); Slovakia (8)	
1.3	case / control status control (0); case (1)	
1.4	sex male (0); female (1); not known (8)	
1.5	date of birth (dd-mm-yy) (not known: all 8)	
1.6	ethnicity Caucasian (1); North African (2); other African (3); Asian (4); Afro-Caribbean (5); other (6); not known (8)	
1.7	subject's marital status single (1); married/cohabiting (2); divorced (3); widow (4); not known (8)	
1.8	Is/was subject right- or lefthanded? righthanded (1); lefthanded (2); not known (8)	
1.9	How many years of full time education did subject complete? (not known: 88)	
1.10	this question only for non-UK cases Has subject lived in or visited the United Kingdom during the period 1980-1996? lived for a period > 6 months (1); lived/visited for a period < 6 months (2); no (3); not known (8)	
1.11	date of interview (dd-mm-yy) (not known: all 8)	

General Information

1.12	place of interview hospital (1); home (2); phone (3); other (4); not known (8)	
1.13	informant spouse (1); self (2); son/daughter (3); nephew/niece/cousin (4); sibling (5); parent (6); other (7); not known (8)	2
z. K	How long has the informant known subject? (in years)	
1.14	interviewer research physician (1); research assistant (2); treating physician (3); nurse (4); other (5); not known (8)	
1.15	sequence of interview before discussion of possible CJD diagnosis with family (1); after discussion(2); not known (8)	

Identification control

controls only

2.1	Identification number of matched case	
2.2	Origin of control hospital control with neurologic disease (1); hospital control without neurologic disease (2); nursing home control with neurologic disease (3);); nursing home control without neurologic disease (4); GP control (5)	
2.3	Controls matched for	
	hospital no (0); yes (1); not known (8)	
2.4	Number of controls refused for matched case (not known; 88)	

Medical Record

cases only

3.1	source neurologist (1); psychiatrist (2); (neuro)pathologist (3); death certificate (4); general practioner (5); EEG department (6); relatives (7); other (8); not known (9)	
3.2	date at notification (dd-mm-yy) (not known: all 8)	
3.3	date at first symptoms (dd-mm-yy) (not known: all 8)	
3.4	date at clinical diagnosis (dd-mm-yy) (not known: all 8)	
3.5	When did patient first seek medical attention? (dd-mm-yy) (not known: all 8)	
3.6	When was patient referred to a neurologist? (dd-mm-yy) (not known: all 8)	
3.7	vital status of case at time of interview dead (1); alive (2); not known (8)	
3.8	duration of illness (in months, defined from first symptoms) (not known 888)	
3.9	date of death (dd-mm-yy) (not known: all 8)	
	Seen alive	Y/n

Clinical Presentation

cases only

Stress-situatic

4.1 <u>neurological signs / symptoms</u>

coding:	no (0) not ki); yes (1); nown (8)	if yes, record date first seen (dd-mm-yy)(not known: all 8)
in _e	rapidly progressive dementia		
×	cerebellar signs		
	visual signs		
	oculomotor signs		
6	pyramidal signs		
	extrapyramidal signs		
	primitive reflexes		
	seizures		
**	myoclonus		
	other involuntary movements		
	headache		
	pain		
	other sensory disturbances		
	vertigo / dizziness		
	pseudobulba <u>r</u> signs	8 4	

	neurogenic muscle wasting		
	akinetic mutism		
4.2	clinical symptoms / signs		
8	gait disturbances		
	speech disturbances		
	visual disturbances		
	forgetfulness		
4.3	psychiatric symptoms/ signs		* *
	clinical depression	è	
	social withdrawal		
ě	low mood and apathy		
	anxiety		
£ .	hallucinations		
	aggression		
4.4	Since the start of the illness, has the patient be by a psychiatrist? no (0); yes (1); not known (8)	en seen	
æ	If yes, record the date of the first consultation (dd-mm-yy) (not known: all 8)		

	Investigations	cases only
		8
5.1	Did the patient undergo an EEG? no (0); yes (1); not known (8)	
	If yes, on how many occasions? (not known: 88)	
	If yes, record date of the last EEG (dd-mm-yy) (not known; all 8)	
5.2	Did the patient recorded an EEG characteristic of CJD (generalized triphasic complexes with frequency about 1/s)? no typical EEG (0); yes, typical EEG (1); no EEG recorded (2); not known (8)	
	If yes, record the date on which the first characteristic EEG was recorded (dd-mm-yy) (not known: all 8)	
5.3	Did the patient have a CT-scan of the brain? no (0); yes (1); not known (8)	
	If yes, on how many occasions? (not known: 88)	
14	If yes, record the date of the last scan (dd-mm-yy) (not known: all 8)	
4.4	Did the collection of the coll	
5.4	Did the patient record an abnormal CT-scan? no (0); yes (1); no CT-scan (2); not known (8)	
s	If yes, record the date on which the first abnormal scan was performed (dd-mm-yy) (not known: all 8)	
	If yes, specify what abnormalities have been observed: no (0); yes (1); not known (8)	

cerebral atrophy

cerebellar atrophy

increase in signal density in thalamus

increase in signal density in (other) basal nuclei

5.5	Did the patient have a MRI scan of the brain? no (0); yes (1); not known (8)	
	If yes, on how many occasions? (not known: 88)	
¥.	If yes, record date of the last scan (dd-mm-yy) (not known: all 8)	
5.6	Did the patient record an abnormal MRI-scan? no (0); yes (1); no MRI scan (2); not known (8)	
	If yes, record the date on which the first abnormal scan was performed (dd-mm-yy) (not known: all 8)	
	If yes, specify what abnormalities were observed: no (0); yes (1); not known (8)	
	cerebral atrophy	
	cerebellar atrophy	
	increase in signal density in thalamus	
	increase in signal density in other basal nuclei	
	22	
5.7	CSF findings	Ð
	protein (mg / dl)	
	glucose (mmol / L)	
	cell count (count / mm ³)	
	14-3-3 (negative (1); equivocal (2); positive (3))	
	NSE (ng / ml)	
	S 100b (ng / ml)	
	tau (pg / ml)	

5.0	results recorded? no (0); yes (1); not known (8)	2
40	If yes, please specify:	normal (0); elevated (1); not known (8) concentration
141	total bilirubin (μMol/L)	
	GOT (IU/L)	
	GPT (IU/L)	
	ALP (IU/L)	
	γGTP (IU/L)	
5.9	Does the patient have any other biochemical/ haematologic abnormalities? no(0); yes (1); not known (8)	
ī	If yes, please descripe the investigations and abnormalities	
	· ·	8
5.10	Did the patient undergo a brain biopsy during life? no (0); yes (1); not known (8)	
e e	If yes, what was the result? no evidence of spongiform change (1); spongiform change without florid plaques (2); spongiform change with florid plaques (3); no biopsy (4); not known (8)	
5.11	Did the patient undergo a tonsil biopsy? no (0); yes (1); not known (8)	
	If yes, what was the result? no evidence of PrP immunostaining (1); equivocal (2); PrP positive (3); no biopsy performed (4); not known (8)	

5.12	Post mortem performed? no (0); yes (1); not known (8)	
9	If yes: nerve cell loss	
No.	gliosis	
	spongiform change	
3	immunohistochemically positive	
	if yes, immunostaining Western blottest	
30	Scrapie Associated Fibrils	
	If yes, diagnosis CJD confirmed? classical CJD (1); nvCJD (2); not known (8)	
5.13	Prion protein gene analysis performed? (chrom. 20) no (0); yes (1); not known (8)	
	If yes, mutation present? no (0); yes (1); not known (8)	
	if yes, specify PRNP mutation	
5.14	polymorphism codon 129 MM (1); MV (2); VV (3); not known (8)	

	Classification	cases only
6.1	How is the patient classified? sporadic (1); familial (2); latrogenic (3); new variant (4); GSS (5)	
6.2	In patients with sporadic CJD, what is the classification according to diagnostic criteria? definite (1); probable (2); possible (3)	
6.3	If familial CJD or GSS, is the diagnosis based on:	
	first degree relative with definite or probable CJD? no relative with CJD (0); relative with definite CJD (1); relative with probable CJD (2); not known (8)	
	disease-specific PRNP mutation? no (0); yes (1); not known (8)	
6.4	At notification to the CJDSU, what was the patient's classification? definite CJD/GSS (1); probable CJD/GSS (2); possible CJD/GSS (3); diagnosis unclear (4); CJD/GSS unlikely (5); definitely not CJD/GSS (6); not known (8)	5 8
	If patient was classified as at least possible CJD/GSS, which category of disease was suspected? sporadic (1); familial (2); latrogenic (3); new variant (4); GSS (5); not known (8)	
6.5	If the patient was seen in live by a member of the CJDSU what was the classification of the patient at the time he/she was first seen by a member of the CJDSU? definite CJD/GSS (1); probable CJD/GSS (2); possible CJD/GSS (3); diagnosis unclear (4); CJD/GSS unlikely (5); definitely not CJD/GSS (6); not seen in live by a member of the CJDSU (7); not known (8)	
	If patient was classified as at least possible CJD/GSS,	

which category of disease was suspected?

GSS (5); not known (8)

6.6

sporadic (1); familial (2); iatrogenic (3); new variant (4);

What was the highest classification on the basis of

clinical information alone (excluding EEG and CSF)? probable CJD/GSS (2); possible CJD/GSS (3); neither probable nor possible CJD/GSS (4); not known (8)

bril			19
Since 1980 has the other than fillings no (0); yes (1); not	(extractions or	dental treatment root canal work)?	
Has the subject e eye operations or admission)? no (0); yes (1); not	stitching of wou	erations, including unds (prior to this	×
If yes, record num and last operation		ns and year of first 8)	ę
	number of operations	year first operation	year last operation
brain		year mst operation	year last operation
spine			
other neurologic			
eye			
ear.			
tonsilectomy			
abdominal			
orthopaedic			
other			

cosm. swg.

7.3	including corneal or bor no (0); yes (1); not known		
	If yes, record number of and last operation (not keep)	of operations and year of first known: 8888)	
		umber of perations year first operation	year last operation
	comea		
	bone marrow		
9	kidney		
	liver		
	other		
	Skin		
7.4	Has subject ever receiv to this admission)? no (0); yes (1); not known	ved a blood transfusion (prior (8)	blooddon o
	If yes, record number of transfusion (not known: 8	f times and year of first and last 3888)	11.
		nber of sfusions year first transfusion	year last transfusion
7.5	Has subject ever receive or immunoglobulin (pr no (0); yes (1); not known		
	If yes, record number of transfusion (not known; 8	f times and year of first and last 3888)	*
50	numb		
	alb/lg transf	fusions year first transfusion	year last transfusion
7.6	of:	dependent diabetes mellitus (2);	
	outer (o), never autilities (TI, HOUNIUMII (O)	

1.1	outpatient because of: bowel disease (1); insulin-dependent diabetes mellitus (2); other (3); never (4); not known (8)	
7.8	Has subject ever visited his/her GP more than 6 times / year because of: bowel disease (1); insulin-dependent diabetes mellitus (2); other (3); never (4); not known (8)	
7.9	Has subject ever been to see a psychiatrist (do not include consultations for current illness) no (0); yes (1); not known (8)	
	If yes, why? depression (1); psychotic disease (2); other (3); not known (8)	
	If yes, record number of years and year of first and last treatment (not known: 8888)	*
	number of treatments year first treatment	year last treatment
	medication	
	ECT	
	hospitalisation	
7.10	Has subject taken any medication regularly, i.e. for a period > 1 months, since 1980 (do not include medications related to current illness) no (0); yes (1); not known (8) prompt for prescription drugs? no (0); yes (1); not known (8)	
京	prompt for hormone therapy or supplements including oral contraceptives and hormone replacement	
	therapy? no (0); yes (1); not known (8)	
	prompt for homeopathic/herbal therapy? no (0); yes (1); not known (8)	
	prompt for eyedrops? no (0); yes (1); not known (8)	
	Summary: has subject been exposed to one of the medications of bovine origin withdrawn in 1990? (see list) no (0); yes (1); not known (8)	
	A Company of the Comp	

7.11	needles? no (0); yes (1); not known (8)	or allergy using	
	If yes, record number of times testing: (not known: 8888)	and year of first and last	
	number of tests	year first test	year last test
7.12	Has subject ever received a tre course of injections (prior to t no (0); yes (1); not known (8)		
	If yes, record name of therapy, first and last treatment (not know		
	name of therapy number of treatments		year last treatment
7.13	Since 1980, has subject been v no (0); yes (1); not known (8)	raccinated?	
	If yes, record type of vaccin, nu and year of first and last vaccin		ä
	type of vaccin number of vacc's	year first vaccination	year last vaccination
7.14	Has subject ever undergone lui electrical tests involving needle no (0); yes (1); not known (8)	•	
đ.	If yes, record number of times a puncture or testing: (not known:		
	number of punctures	year first puncture	year last puncture

7.15	no (0); yes (1); not known (8)		
	If yes, record number of times and year of first ar acupuncture: (not known: 8888)	nd last	
*	number of times year first acuj	puncture year last acupunc	ture
7.16	Has subject ever used recreational drugs, for example cannabis, ecstasy, heroin or crack? no (0); yes (1); not known (8)		
	If yes, record route intravenous (1); oral (2); inhaling (3); not known (8)		
10	If yes, record number of years and year of first an use: (not known: 8888)	nd last	
	number of year first use	year last use	
7.17	has subject ever been tattoed? no (0); yes (1); not known (8)		
in the	If yes, record number of times and year of first an tattoe: (not known: 8888)	d last	104
	number of tattoes year first tatto	e year last tattoe	
7.18	Has subject ever undergone ear or body piercin no (0); yes (1); not known (8)	g?	5
	If yes, record number of times and year of first and piercing: (not known: 8888)	d last	
	number of piercings year first pierc	cing year last piercing	

Family history

8.1

	CJD		OTHER DE	OTHER DEMENTIA'S	
	no (0) yes, definite(1) yes, probable (2) not known (8)	age at onset	no (0); yes (1); not known (8)	age at onset	
father				20	
mother					
grandmother father	٠				
grandfather father					
grandmother mother		285-20-30			
grandfather mother					
sibling 1					
sibling 2				NEC.	
sibling 3					
sibling 4					
sibling 5					
sibling 6					
sibling 7					
sibling 8			E C		
sibling 9		82			
sibling 10					
sibling 11			***************************************		
sibling 12					

8.2	Has subject had social contact, through family, friends or work, with someone else who developed CJD? no (0); yes (1); not known (8)	
	If yes, record the person's appoximate date of illness (dd-mm-yy) (not known: all 8)	
	Confirmation of social contact with case of CJD definite (1); probable (2); possible (3); unable to concern (4); not a case (5); not known (8)	

Die	etary hi	story				- Par			
year	subject ever	and the same resemble	etarian fo	or a peri	od of 1	25			
no (0); yes (1); not	known (8)	10 N						
befor	s, during who re 1980 (1); 19 -1996 (4); not e than one of	980-1985 (2); known (8)		0 (3);					
If ye	s, for how ma	any years? (not known	: 88) =	121				
time	s, did (s)he e ? i); yes (1); not		or fish at	all duri	ng this				
cat	e the beginn or dog food'); yes (1); not	?	has subje	ect ever	eaten			a a	
	s, how? d (1); dried <u>(</u> 2)); tinned and	dried (3); n	ot know	1 (8)			5	
well o	does/did sul done (1); med teak (5); not k	ium (2); medi							
since	each of the for the beginni shown on the	ng of 1980?							
		24		8		freq.	beef (3); h veal (e code orig (1); pork (2) orse (4); ch 6); venison n (8) <i>more</i> o ble	; lamb icken (5) (7); not
		5			brains).	9.34		
		N.			eyes	2			
				B 16	tripe	191			
			٠	,;	liver				
			n .			240			

Goat (meat / ch	eese)	
kidneys		
sweetbreads (pancreas)		
readymade burgers		
readymade meat pies such as pork, veal and ham, steak and kidney, chicken and mushroom		
meat sausages		
black pudding		
liver sausage or paté		
steak tartare (raw minced steak with raw egg)/carpaccio		
cheese		
roast lamb, lamb chops, lamb stew		
haggis		*
roast pork, ham, bacon		
faggots		
roast beef, steak, beef stew, minced beef		ψ.
veal		*
venison		
chicken		
cow's milk drinks milk/eats breakfast cereal with milk (1); only in tea/coffee (2); no (3); not known (8)		

Exposure to animals

		27	
10.1	Since the beginning of 1980 home with:	, has subject shared a	
		cats	
* *		no (0); yes (1) not known (8) dogs	
	20	no (0); yes (1) not known (8)	
		ferrets	
		no (0); yes (1) not known (8)	
10.2	Since the beginning of 1980 stayed for more than one we lived or worked (1); stayed (2);	eek on a farm?	
			e to
	If yes, did subject work or he	elp with:	
		cattle	
	2	no (0); yes (1) not known (8)	
	Ę.	sheep no (0); yes (1) not known (8)	
	Ø	goats no (0); yes (1) not known (8)	
	*	pigs no (0); yes (1) not known (8)	
		chickens no (0); yes (1) not known (8)	
		mink no (0); yes (1) not known (8)	
	42		781
	If yes, did subject participate	in deer	
	treating cattle for	r warble fly (with phosmet)	
¥.	treating cattle for	no (0); yes (1) not known (8)	
		dipping sheep	
		no (0); yes (1) not known (8)	-
		crop spraying	3 3
		no (0); yes (1) not known (8)	
10.3	Since the beginning of 1980, the following:	, has subject used any of	2
		w	
	× v	bonemeal	
	= **	no (0); yes (1) not known (8) hoof and horn	
		no (0); yes (1) not known (8)	
		dried blood	
±I		no (0); yes (1) not known (8)	
*		manure	
		no (0); yes (1) not known (8)	

0.4	meat or bon Ask responder	pes/did subject cut or c es, in his/her work or a nt to indicate the most app own on the flash card.	t home?		* ×	
0.5	Has subject of at school? no (0); yes (1)	ever dissected animal not known (8)	eyes, for example		2 2 a	ж
	4			× "		
			a *	a	et.	
	*				i i	
	×		a)			
		0 0		E A		
	WI.					

Occupation	
	The state of the s

from last onset (not	number o elapsed fi work to or applicable	ever (1); never (2); not known (8)	coding::	1001
			Has the subject ever worked in farming, the meat industry, the pharmaceutical industry or in a hospital	11.1
			Has subject worked in the following areas: medical/paramedical/nursing/dentistry	11.2
			animal laboratories	
			pharmaceutical laboratories	
			other research laboratories	50
			animal farming/veterinary medicine	
			meat industry (butcher's/abattoir/rendering plants etc)	
			catering industry	
		Y/n	other occupation involving animal products (e.g. leather worker) hunting Has subject's partner worked in the following areas:	11.3
			medical/paramedical/nursing/dentistry	
¥			animal laboratories	
			pharmaceutical laboratories	
			other research laboratories	

	animal farming/veterinary medicine		
	meat industry (butcher's/abattoir/rendering plants etc)		
38	catering industry		
11.4	other occupation involving animal products (e.g. leather worker) hunting Was subject's parents' most recent occupation in any of the following areas:	/n	
	medical/paramedical/nursing/dentistry		
	animal laboratories		
	pharmaceutical laboratories		
	other research laboratories		
	animal farming/veterinary medicine		
	meat industry (butcher's/abattoir/rendering plants etc)		
	catering industry		
	other occupation involving animal products (e.g. leather worker)		
	hunting 4	In	

Residential History:

s s

r n

*

Coding for food frequencies

Response	Code
	a
Never	1
less than once per year	2
about once per year	. 3
several times per year	4
about once per month	5
about once per week	6
several times per week	7
several times per day	8

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Coding for food frequencies 2				