Republic of Lebanon
Ministry of Public Health
Directorate of Prevention

Epidemiological Surveillance Unit

الجمهورية اللبنانية
مكتب وزير الدولة لشؤون التنمية الإدارية
مركز مشاريع ودراسات القطاع العام

Rapid Study on Prevalence of Creutzfeld Jakob Disease in Lebanon 2001

Republic of Lebanon
Office of the Minister of State for Administrative Reform
Center for Public Sector Projects and Studies
(C.P.S.P.S.)

Prepared by:
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REPUBLIC of LEBANON
Ministry of Public Health
Directorate of Prevention
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Rapid Study on Prevalence of Creutzfeld-Jakob Disease in Lebanon

MOH, WHO and the Lebanese Neurologists and Psychiatrists.

1- Context

Since year 1996, awareness was raised in Europe concerning the Transmissible Spongiform Encephalopthy in Humans. In Humans, 4 etiologies are documented: the sporadic form (for more than 50 years old), the genetic form (as GSS, FFI...), the iatrogenic form and the new variant of Creutzfeld-Jakob Disease. The last one has hypothetical relations with the Bovine Spongiform Encephalopthy. Since 1998, Creutzfeld-Jakob Disease has been included among the mandatory reportable communicable diseases in Lebanon.

2- Objective

The aim of the rapid assessment is to estimate the prevalence of the Human Spongiform Encephalopthy or Creutzfeld-Jakob Disease in Lebanon during the last 5 years.

It is well known that the average prevalence of sporadic cases in the world is at least one case per one million habitants by year. In Lebanon we should expect 4 cases by year (the Lebanese population is estimated to 4 millions habitants). Previous to year 2001, no cases were notified to the Ministry of Health. In year 2001, 3 cases were reported to the Ministry in Lebanon.

3- Material and Methods

A Circular was issued by the Ministry of Health, number 54 dated on 20 august 2001, asked all Lebanese Neurologists and Psychiatrists to report any case of Creutzfeld-Jakob Disease, using the specific questionnaire related to the study and to send it to the Epidemiological Surveillance Unit (see Annex).

The questionnaire was standardized. It includes all case definitions adopted by the WHO for all Human Spongiform Encephalopthies (see Annex). Only the diagnosis techniques available in Lebanon have been considered. There are:

- The case definition for possible sporadic case of CJD: Progressive dementia with at least two out of:
  - -myoclonus
  - -visual or cerebellar disturbance
  - -pyramidal/extra-pyramidal dysfunction
  - -akinetic mustism
  - **with or without** periodic tracing on EEG
  - **with** a clinical duration to death <2 years
- The case definition of iatrogenic case of CJD: Progressive cerebella syndrome following the use of human cadaveric pituitary hormone or progressive dementia following neurosurgery (corneal transplant, dura mater graft, stereotaxic EEG...)
- The case definition for familial case of CJD: Progressive dementia with a similar case for the father or the mother
- The case definition for suspected new variant case of CJD: Progressive neuropsychiatric disorder with at least four out of:
  - -early persitent parasthesisa /dysaesthesia
  - - ataxia
  - -chorea/dystonia/myoclonus
  - -dementia
  - -akinetic mustism
- The case definition for definite case of CJD: Spongiform encephalopathy on the neuropathology.

The period asked for was the years 1997, 1998, 1999, 2000 and 2001.

. Physician's names and addresses were provided by the two Orders of Physicians of Lebanon and of North Lebanon according to their listings of year 2000.

Physicians were contacted by phone; the circular and questionnaire were sent by fax from the Epidemiological Surveillance Unit. The filled questionnaire were received by fax to the Unit. The study was conducted during the third and fourth quarters of year 2001.

Data entry and descriptive analysis were done using the Epiinfo software.

4- Results

- **Response rate**: 144 Neurologists and Psychiatrists were registered at the Orders of Physians in Lebanon. 75 % of them have been contacted. Among the contacted physicians, 62 % of Neurologists and 37 % of Psychiatrists did participate and reply to the study.

Table (1): Number of Physicians included, contacted and responded for the study

	Nb registered to the Physicians Orders	Nb contacted	Nb responded
Neurologists	96	73	45
Psychiatrists	48	35	13
Total	144	108	58

Source: Rapid assessment of CJD in Lebanon – MOH – WHO - 2001

Table (2): Percentages of Physicians included, contacted and responded for the study

	Nb registered to the Physicians Orders	% contacted / registered	% responded / contacted
Neurologists	96	76 %	62 %
Psychiatrists	48	73 %	37 %
Total	144	75 %	54 %

Source: Rapid assessment of CJD in Lebanon – MOH – WHO - 2001

- **Reasons of no contact**: Among the 36 remained physicians for whom we failed to contact them, 13 over 36 (36 %) were retired, abroad or dead.

Table (3): Reasons of no contact

	Retirement	Emigration	Death	Other	**Total**
Neurologists	1	4	0	18	23
Psychiatrists	2	4	2	5	13
Total	3	8	2	23	36

Source: Rapid assessment of CJD in Lebanon – MOH – WHO - 2001

- **Case finding**: 8 neurologists did report 10 suspected cases during the past 5 years, which is equivalent to an overall of 18 % of the Neurologists. The Psychiatrists reported no case.

Table (4): Number of Physicians reporting CJD and number of cases

	Nb of Physicians with cases	Nb of cases
Neurologists	8	10
Psychiatrists	0	0
Total	8	10

Source: Rapid assessment of CJD in Lebanon – MOH – WHO - 2001

Table (5): Percentage of Physicians who reported CJD

	% Physicians with cases / responded
Neurologists	18 % (8/45)
Psychiatrists	0 % (0/13)
Total	14 % (8/58)

Source: Rapid assessment of CJD in Lebanon – MOH – WHO - 2001

- **The cases**: 10 case of CJD were reported from the previous years. 5 were diagnosed in 2001, 2 in 2000, 2 in 1999 and 1 in 1998. No case was reported in 1997. According to the clinical classification, 8 were sporadic cases of CJD, 1 familial and 1 case of new variant of CJD. Also some physicians reported Alzheimer and vascular dementia cases that were similar to CJD case definitions.

Table (6): Number of reported cases of CJD

Case Definition		Years						Total
		1997	1998	1999	2000	2001	UN	
Possible Sporadic	Progressive dementia with at least two out of: -myoclonus -visual or cerebellar disturbance -pyramidal/extra-pyramidal dysfunction -akinetic mustism **with or without** periodic tracing on EEG **with** a clinical duration to death <2 years		1	2	1	4		8
Iatrogenic	Progressive cerebella syndrome following the use of human cadaveric pituitary hormone							0
	Progressive dementia following neurosurgery (corneal transplant, dura mater graft, stereotaxic EEG...)							0
Familial	Progressive dementia with a similar case for the father or the mother				1		* Alzheimer	1
Suspected new variant	Progressive neuropsychiatric disorder with at least four out of: -early persitent parasthesisa /dysaesthesia - ataxia -chorea/dystonia/myoclonus -dementia -akinetic mustism					1	** Vascular dementia	1
Definite	Spongiform encephalopathy on the neuropathology							0
Total		0	1	2	2	5		10

Notes: * Alzheimer cases (1986-1991), ** Vascular dementia cases

Source: Rapid assessment of CJD in Lebanon – MOH – WHO - 2001

5- Discussion

Based on the clinical definitions of CJD and the physician's memory, more reliable information is obtained for the last year 2001.

In 2001, 3 cases of CJD were reported to the Ministry of Health. The 3 cases were:

- The first case was diagnosed clinically and classified by one of the Lebanese Neurologist expert as a sporadic case of CJD. This case was reported in the study as sporadic case.
- The second case was reported as suspected new variant CJD, the patient died at 30 years old. However the clinical history had began 4 years before and a medical records review by the CJD Surveillance Unit in UK discarded the case as being CJD. This case was reported in the study as sporadic case.
- The third case was also reported as suspected new variant of CJD, the patient died at 48 years old. The autopsy was performed. The immunocytochemistry for PrP (done at the National CJD Surveillance Unit in UK) was negative and the case was discarded as being CJD. This case was reported in the study as new variant.

6- Conclusion

During this rapid assessment of CJD in Lebanon, 10 cases were found. However medical records review by experts is necessary to classify the cases. Contacts are made between the Ministry of health and the Lebanese Society of Neurology to designate a national committee who will responsible for reviewing each case found or reported as suspected case of CJD and for establishing national guidelines on diagnosis and investigation procedures for CJD.

On the other hand the Ministry of Health should assure the feed back for physicians on all reported cases of CJD. A summary of this rapid assessment will be send to all Lebanese Neurologists and Psychiatrists (see Annex).

7- Thanks

We thank the neurologists who were willing to participate to this study: DRS ABBAS SERHAN, ABED EL RASOUL SAYEG, ABED EL REHMEN SAMRA, ANTOINE AWAD, BASSEM YAMOUT, DANIEL TABIB, ELIE ASSAF, FADI ABOU MRAD, FAYEK YOUNESS, GEORGES HANNA, GHADA SABBAGH, HANNA MATTAR, HASSAN HAMOUD, HASSAN TRABOLSI, JAMAL HNEINI, JEAN TAMRAZ, KAMAL KALLAB, KASSEM KHALAF, KHALED MASRI, KHODOR SEKLAWI, KIVORK AGOPIAN, MAROUN MOUSSA, MIKHAEL FERZLI, MOUHAMAD KASSEM, MOUNIR KHOURY, MOUNIR RAHME, NAGI RIYACHI, RAJA DIBO, RAYMOND CHIMALI, REDA ABOU CHAHIN, RENE GEMAYEL, RYAD KHALIFEH, SALIM ATROUNI, SALIM MASRI, SAMI TOHME, SAMIR AMAR, SAMIR ATWI, SHAKER KHAMISS, SHAWI YOUSSEF, SHAWKAT BAAINE, SOUHEIL GBEILY, WAEL MASRI, ZAKHIA SALIBA, ZIAD RIFAII, ZOUHEIR MOUSSA.

We thank the psychiatrists who were willing to participate to this study: DRS ADEL STEPHAN AKEL, AMER HAYDAR, ANDRE PHILIP BALADI, ARTINE JOSEPH CONIAN, CHARLES BADOURA, ELIE GEORGES KARAM, ELIAS CHEDID, ELIAS SASSINE, FRANCOISE GHOUSOUB, GEORGES SAMIR GAMOUS, JEAN ANTOINE FIYAD, JOSIANE MAROUN MADI, NIZAR HALABI.

Questionnaire

الجمهورية اللبنانية
وزارة الصحة العامة
مديرية الوقاية الصحية
وحدة الترصد الوبائي

دراسة وطنية خاصة بمرض الاعتلال الاسفنجي الدماغي عند الانسان في لبنان

Human Spongiform Encephalopathy - Creutzfeldt Jakob Disease

Physician name: ..... Specialty: ..... Number Order: .....

Address: .....

Phone number: .....

Fax number: .....

During the 5 past years (1997, 1998, 1999, 2000 and 2001), have you diagnosed a suspected or confirmed case of Creutzfeld-Jakob Disease in Lebanon ?

Name (year)		
.....	Yes / No	Progressive dementia with at least two out of : -myoclonus -visual or cerebellar disturbance -pyramidal/extrapyramidal dysfunction -akinetic mustism **with or without** periodic tracing on EEG **with** a clinical duration to death <2years
.....	Yes / No	Progressive cerebellar syndrome following the use of human cadaveric pituitary hormone
.....	Yes / No	Progressive dementia following neurosurgery (corneal transplant, dura mater graft, stereotaxic EEG...)
.....	Yes / No	Progressive dementia with a similar case for the father or the mother
.....	Yes / No	Progressive neuropsychiatric disorder with at least four out of: -early persitent parasthesisa /dysaesthesia - ataxia -chorea/dystonia/myoclonus -dementia -akinetic mustism
.....	Yes / No	Spongiform encephalopathy on the neuropathology

Date : Physician signature:

Please send this questionnaire to the Epidemiological Surveillance Unit – Tel : (01) 614194, Fax : (01) 610920 or (01) 615759

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LE DIRECTEUR GÉNÉRAL

الجمهورية اللبنانية
وزارة الصحة العامة
المدير العام

رقم المحفوظات: ١/١ – ١/٢٠٢
بيروت في ٢٠ آب ٢٠٠١

تعميم رقم ٥٤

دراسة وطنية خاصة بمرض الاعتلال الاسفنجي الدماغي عند الإنسان في لبنان

في إطار ترصد حالات الاعتلال الاسفنجي الدماغي عند الإنسان في لبنان –
Human Spongiform Encephalopathy – Creutzfeldt Jakob Disease

تقوم وزارة الصحة العامة ، بإجراء دراسة وطنية سريعة ، حيث يسأل كل طبيب من ذوي اختصاص الأمراض العصبية وذوي الاختصاص الأمراض النفسية ، إذا عاين مريضا مشتبه أو مؤكد انه مصاب بمرض الاعتلال الإسفنجي الدماغي ، خلال الخمس السنوات الأخيرة أي منذ ١٩٩٧ حتى الآن.

لذا نتمنى على جميع الأطباء المعنيين ، التعاون مح وحدة الترصد الوبائي التابعة لمديرية الوقاية الصحية وذلك عبر ملاء الاستمارة المرفقة وإرسالها إلى الوزارة عبر الفاكس : ٠١-٦١٠٩٢٠ أو ٠١-٦١٥٧٥٩.

و على ضوء معطيات الدراسة ، ستدرس الملفات المشتبهه وتنشئ قاعدة معلومات طبية وطنية خاصة في مرض الاعتلال الإسفنجي الدماغي عند الإنسان في لبنان .

كما نؤكد أن المعلومات ستظل تحت حماية السرية الطبية و السرية المهنية. و نشكركم لتعاونكم.

عن مدير عام وزارة الصحة العامة
رئيس مصلحة الديوان
حكمت اسعد

الجمهورية اللبنانية
وزارة الصحة العامة
مديرية الوقاية الصحية
وحدة الترصد الوبائي

دراسة وطنية خاصة بمرض الاعتلال الاسفنجي الدماغي عند الانسان في لبنان
Human Spongiform Encephalopathy - Creutzfeldt Jakob Disease

اسم الطبيب: ..... الاختصاص: ..... رقم النقابة: .....
عنوان العمل: .....
رقم الهاتف: .....
رقم فاكس العمل: .....

خلال السنوات الخمسة الاخيرة - أي ١٩٩٧ ، ١٩٩٨ ، ١٩٩٩ ، ٢٠٠٠ و ٢٠٠١ - هل عاينت مريضا **مشتبه** او مثبت ان يكون مصابا بمرض الاعتلال الاسفنجي الدماغي عند الانسان ، في لبنان ؟

الاسم (و السنة)		
	نعم / كلا	Progressive dementia with at least two out of : -myoclonus -visual or cerebellar disturbance -pyramidal/extrapyramidal dysfunction -akinetic mustism **with or without** periodic tracing on EEG **with** a clinical duration to death <2years
	نعم / كلا	Progressive cerebellar syndrome following the use of human cadaveric pituitary hormone
	نعم / كلا	Progressive dementia following neurosurgery (corneal transplant, dura mater graft, stereotaxic EEG...)
	نعم / كلا	Progressive dementia with a similar case for the father or the mother
	نعم / كلا	Progressive neuropsychiatric disorder with at least four out of: -early persitent parasthesisa /dysaesthesia - ataxia -chorea/dystonia/myoclonus -dementia -akinetic mustism
	نعم / كلا	Spongiform encephalopathy on the neuropathology

التاريخ : توقيع الطبيب:

الرجاء ارسال الاستمارة معبئة الى وحدة / الترصد الوبائي - رقم الهاتف : 01-614194/5/6 - *رقم الفاكس :* 01-610920 or 01-615759

REPUBLIC of LEBANON

Rapid Study on Prevalence of Creutzfeld-Jakob Disease in Lebanon

Ministry of Public Health, WHO, Lebanese Neurologists and Psychiatrists.

1. **Context:** Since year 1996, awareness was raised in the world concerning the Creutzfeld-Jakob Disease CJD. 4 etiologies are documented: the sporadic form (for more than 50 years old), the genetic form, the iatrogenic form and the new variant of CJD. Since 1998, CJD has been included among the mandatory reportable communicable diseases in Lebanon.

2. **Objective:** The aim of the rapid assessment is to estimate the prevalence of the CJD in Lebanon during the last 5 years. According to average prevalence of sporadic cases in the world (1 case/1 million habitants), it is expected to have 4 cases by year.

3. **Material and Methods:** A Circular was issued by the Ministry of Health, number 54 dated on 20 august 2001, asked all Lebanese Neurologists and Psychiatrists to report any case of CJD, using the specific questionnaire related to the study and to send it to the Epidemiological Surveillance Unit. The questionnaire was standardized. It includes all case definitions adopted by WHO for all CJD. Only the diagnosis techniques available in Lebanon have been considered. The case definitions include the definition for the possible sporadic case of CJD, the iatrogenic case of CJD, the familial case of CJD, the suspected new variant case of CJD and the definite case of CJD.
The period asked for was the years 1997, 1998, 1999, 2000 and 2001. Physician's names and addresses were provided by the two Orders of Physicians of Lebanon and of North Lebanon according to the listings of year 2000. Physicians were contacted by phone and fax during the third and fourth quarters of year 2001. Data entry and descriptive analysis were done using the Epiinfo.

4. **Results:** 144 Neurologists and Psychiatrists were registered at the Orders of Physicians in Lebanon. 75 % (108/144) of them have been contacted. Among the contacted physicians, 54 % (58/108) did reply to the study. Among the remained 36 physicians for whom we failed to contact them, 36 % (13/36) were retired, abroad or dead. 8 neurologists did report 10 suspected cases during the past 5 years, which shows an overall of 18 % (8/45) for the Neurologists. The Psychiatrists reported no case.

Table: Number of reported cases of CJD

Case Definition		Years						Total
		1997	1998	1999	2000	2001	UN	
Possible Sporadic	Progressive dementia with at least two out of: -visual or cerebellar disturbance -akinetic mustism -pyramidal/extra-pyramidal dysfunction -myoclonus **with or without** periodic tracing on EEG **with** a clinical duration to death <2 years		1	2	1	4		8
Iatrogenic	Progressive cerebella syndrome following the use of human cadaveric pituitary hormone							0
	Progressive dementia following neurosurgery (corneal transplant, dura mater graft, stereotaxic EEG...)							0
Familial	Progressive dementia with a similar case for the father or the mother				1		* Alzheimer	1
Suspected new variant	Progressive neuropsychiatric disorder with at least four out of: -early persitent parasthesisa /dysaesthesia -chorea/dystonia/myoclonus -ataxia -akinetic mustism -dementia					1	** Vascular dementia	1
Definite	Spongiform encephalopathy on the neuropathology							0
Total		0	1	2	2	5		10

Source: Rapid assessment of CJD in Lebanon – MOH – WHO - 2001

1- **Discussion:** Based on the clinical definitions of CJD and the physician's memory, more reliable information is obtained for the last year 2001. In 2001, 3 cases of CJD were reported to the Ministry of Health, there were one sporadic case of CJD and two suspected new variant that have been both discarded: one of them had a clinical history lasting for more than 4 years and the second case had negative immunocytochemistry for PrP (performed after autopsy). The last case is the new variant CJD case found in the study for year 2001.

2- **Conclusion:** Medical records review by experts is necessary to classify the cases. A national committee is needed for reviewing each case reported as suspected case of CJD and for establishing national guidelines on diagnosis and investigation procedures for CJD. On the other hand the Ministry of Health should assure the feed back for physicians on reported cases of CJD.

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