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Targeted screening of high-risk cattle populations for BSE to augment mandatory reporting of clinical suspects

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Abstract

In Switzerland, the first case of bovine spongiform encephalopathy (BSE) was diagnosed in November 1990. Case numbers peaked in 1995, with a total of 352 BSE cases identified by 30 April 2000. Reporting of clinically suspect cattle is currently the most commonly used method world-wide to detect BSE cases. The effectiveness of mandatory reporting depends on a variety of factors; for other diseases passive surveillance underestimates the incidence of clinical cases. The efficiency of passive surveillance systems for BSE will remain unknown until screening tests able to identify clinically affected cattle have been applied in several countries. This paper provides the first detailed description of a targeted screening programme for BSE. Two populations of cows >24 months of age were included in the targeted screening: (i) cows found dead or culled on site where the carcass was submitted to rendering (fallen stock) and (ii) cows with health-related problems unfit for routine slaughter that were slaughtered under emergency procedures (emergency slaughter). Between 1992 and 1999, on average 81 clinical BSE suspects per year were reported to the veterinary authorities (passive surveillance), of which 43% were confirmed with BSE. A total of 30 clinical cases were captured by passive surveillance and an additional 20 BSE cases detected by targeted screening between May 1999 and April 2000. The odds of finding a BSE case was 49 times higher in the fallen stock and 58 times higher in emergency-slaughtered cattle when compared to passive surveillance. The targeted screening of fallen stock and emergency-slaughtered cattle considerably increased the number of detected cases in this 12-month period. Targeted-screening cases were on average 4 months younger than the clinical suspect cases.

In conclusion, post-mortem testing of fallen stock and emergency-slaughtered cows >24 months for BSE is an important active surveillance element within a total surveillance system that

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principally is based on mandatory reporting of clinical suspect cases. Without ante-mortem screening tests to detect BSE-infected cattle during the incubation period, a combination of effectively functioning passive and active BSE surveillance strategies might be the only approach to assess the BSE situation reliably in a given country or region — and it is necessary to substantiate claims of freedom from the disease.

Keywords: Bovine spongiform encephalopathy; Cattle; Surveillance; Epidemiology; Screening; Mandatory reporting

1. Introduction

In Switzerland, the first case of bovine spongiform encephalopathy (BSE) was diagnosed in November 1990 (Cachin et al., 1991). Reporting of all cattle suspected of clinical BSE and subsequent laboratory examination became mandatory immediately thereafter. The disease was introduced to the Swiss cattle population most likely as a result of indirect importation of meat-and-bone meal (MBM) originating from Great Britain (Hörnlimann et al., 1994, 1996). The Swiss BSE epidemic peaked in 1995 and then started to decline with a total of 352 identified cases by 30 April 2000. Monthly case updates are available on the internet pages of the Swiss Federal Veterinary Office (http://www.bvet.admin.ch).

Mandatory reporting and investigation of clinically suspect cattle (in the context of this paper referred to as "passive surveillance") is the method used by most countries to detect cases of BSE in the national herd. The effectiveness of a passive surveillance system can be expressed as the number of animals identified, reported and diagnosed, divided by the (yet unknown) number of infected animals expressing sufficient clinical signs of disease to meet the criteria for suspicion in the population under surveillance. This effectiveness depends on a variety of factors including the appropriateness of the case definition, the variability of clinical signs, the disease awareness of the farmers and veterinary practitioners, the quality of the ante-mortem slaughter inspection, consequences of reporting diseased animals (such as the loss of the entire herd), the compensation offered, and the properties of the diagnostic tools involved. For a variety of diseases in animals and humans passive surveillance underestimates the incidence of clinical cases (Lilienfeld and Stolley, 1994; Kelsey et al., 1986; Martin et al., 1987; Toma et al., 1999), and it is likely that the degree of underreporting will vary between countries, regions or age groups (Cohen, 2000). For scrapie in sheep and goats, underestimation levels between 30 and 87% have been reported (Schreuder et al., 1993; Hoinville et al., 1999; Baumgarten et al., 2000), and low case detection and reporting risks for BSE have been discussed in recent modelling approaches for the BSE epidemic in UK and Switzerland (Dealler and Kent, 1995; Cohen and Valleron, 1999; Doherr et al., 1999a; Donnelly and Ferguson, 2000). The true efficiency of mandatory reporting of clinical BSE suspects is — and will remain — unknown until screening tests able to identify clinically affected cattle reliably (Schaller et al., 1999; Moynagh and Schimmel, 1999) have been applied in several countries on clearly defined sub-populations that have a higher incidence of detectable BSE cases than the general cattle population. With data only from passive surveillance systems, a meaningful comparison of BSE incidence figures between countries is difficult — if not impossible.

One of the EU-validated BSE screening tests, a Western Blot (PWB) developed by the Prionics AG, Zürich/Switzerland for post-mortem detection of the protease-resistant core of the BSE-specific prion protein (PrP^{Sc}) in cattle brain tissue (Schaller et al., 1999) was used in Switzerland to screen defined target populations for BSE. In parallel, efforts to heighten disease awareness for BSE to improve the passive surveillance system were increased. Preliminary results of the first 9 months of active surveillance have been presented before (Doherr et al., 1999b, 2000; Heim et al., 1999). The objective of this paper is to provide the first detailed description of the targeted screening programme for BSE on emergency-slaughtered cows and fallen stock begun in Switzerland in January 1999 and to discuss the results of the targeted screening and the ongoing passive BSE surveillance in the context of detection effectiveness.

2. Materials and methods

2.1. Definitions

In this paper, passive-surveillance cases are cattle that were reported to the veterinary authorities as clinical BSE suspects and confirmed by immunohistochemistry (IHC) in the Swiss National Reference Laboratory for BSE. The clinical case incidence is defined as the number of confirmed BSE cases among the reported clinical suspects in a given period of time divided by the average population at risk during this time. Cattle from the screened target populations with detectable levels of PrP^{Sc} in brain tissue in the PWB and in IHC are referred to as "targeted-screening cases". These cases fall into two categories: (i) cows that were removed from the population for health-related reasons and that were slaughtered sick (emergency slaughter) and (ii) cows that were found dead or culled for health-related or age-related problems where the carcass was disposed off (fallen stock). The targeted screening incidence is defined as the number of detected BSE cases in the respective target population for screening over a given period of time divided by the total number of cattle from this target population that were screened during this time.

2.2. Population at risk and BSE cases used

The total Swiss cattle population in 1996 was composed of approximately 1.7 million animals, 0.9 million of which were adult female cattle designated for breeding and dairy production. Cows in this population are typically the only cattle to reach a productive age that is beyond the observed youngest age of a bovid with clinical BSE in Switzerland (Doherr et al., 1999a). This was considered to be the population under passive surveillance.

In the context of this work, all Swiss BSE cases detected by passive surveillance (mandatory reporting of clinical suspects) between 1990 and 30 April 2000 (320), and all cases detected by screening of emergency slaughter cattle and fallen stock until 30 April

2000 were used. BSE cases diagnosed in cows that were tested in the context of the BSE-related herd, progeny and cohort culling schemes (Schaller et al., 1999) as well as three cases detected in a sample of routinely slaughtered cattle between June 1999 and October 1999 (Doherr et al., 1999b) were excluded from the analysis. As temporal reference points for the cases, the date of birth and the date of death or culling were used.

2.3. Passive BSE surveillance

Prior to December 1990, cattle brains submitted for neuropathological diagnosis to the Institute of Animal Neurology (ITN), University of Bern, were also screened for lesions consistent with BSE. Since 1 December 1990, all cattle for which BSE could not be ruled out on clinical grounds have to be reported to the Swiss cantonal veterinarians as "clinical suspect cases". These veterinarians are responsible for culling, compensation, and forwarding brain samples to the ITN (now formally the Swiss Reference Laboratory for BSE) for diagnosis. Confirmation of BSE until 1993 was based on the presence of typical lesions. Since 1994, the IHC to detect PrP^{Sc} in the obex region of the brain of suspected cases was added as a diagnostic tool, and suspect cases with distinct histological lesions or positive PrP^{Sc} staining are classified as BSE. The examination is free of charge, and the owners are compensated for at least 90% of the market value of the suspect cases. Disease awareness was maintained by regular publications in industry journals, the bi-weekly official bulletin of the Swiss Federal Veterinary Office (FVO), by presentations to the cantonal veterinarians, and by continued education workshops for veterinary practitioners. In 1999, several articles and announcements in the bulletin of the FVO were launched to describe the targeted screening and the screening results were presented on the internet page of the FVO. In addition, two continuing education workshops on neurological diseases in ruminants were organised for large-animal practitioners; these workshops stressed the issue of identifying and reporting clinical BSE suspects.

2.4. Screening of target populations (active BSE surveillance)

In experimentally infected cattle, an increase in PrP^{Sc} in certain brain regions to IHC detectable levels occurs shortly before or around onset of clinical disease (Wells et al., 1998). The target population for testing — due to the assumed test characteristics — was therefore restricted to cattle old enough to show clinical BSE and which were removed from the national herd (preferably with health-related problems). At the end of 1998, the previously defined target sub-populations of cows >2 years were selected. It was assumed that these two target populations could contain a sizeable proportion of detectable BSE-infected animals that were missed in the existing passive surveillance. Each year, >9000 adult cows were thought to be subject to emergency slaughter or to leave the population as fallen stock — but at the end of 1998 reliable estimates were available neither for this number nor for the expected incidence of detectable BSE cases in the two populations. A decision therefore was made to examine all adult cows in the two groups.

2.4.1. Fallen stock collection and brain sampling

In Switzerland, all animal carcasses are collected at regional centres and transported to one rendering plant licensed for fallen stock and high-risk material. All meat-and-bone meal produced in this plant subsequently is incinerated. Physical appearance and teething status was used as a proxy for age determination. Since January 1999, collection-truck drivers specifically are instructed to tag collected cow carcasses with a clearly visible BSE ear tag if they show at least two permanent incisors or a well-developed udder. This definition is used to include all cows that were at least 24 months of age. In addition, the drivers are asked to complete a form with information on the owner of the tagged cows and identification numbers (BSE tag, regular ear tags) of the latter. At the central rendering plant, cow carcasses with BSE ear tags are separated from the other carcasses and the veterinarian takes brain-stem samples on site. For clinical-suspect cases, the skull is opened for extraction of the whole brain including brain stem to allow a complete neuropathological examination. In most of the other cases, brain-stem samples are collected with a sharp spoon through the *Foramen magnum.* These samples (accompanied by the forms) are shipped to the Prionics AG in Zürich (to which the testing has been outsourced) for examination with the PWB.

2.4.2. Brain sampling from emergency-slaughtered cows

Veterinarians and meat inspectors were provided with guidelines and the instruments (sharp spoons) necessary to collect brain-stem samples from all adult emergency slaughtered cows through the *Foramen magnum.* Age determination for animals for which the actual birth date was unavailable was done as described for fallen stock. All brain-stem samples are shipped in provided containers by express mail to the Prionics AG for examination in the PWB. The carcasses of sampled cows and their slaughter offal are held until the test results are available. The Prionics AG is required to fax the results back to the slaughter plants the morning after the samples arrived (so that the carcasses and offal can be either released or destroyed).

2.4.3. Testing of brain-stem samples

Sample preparation and Western-Blot detection of the protease-resistant core of PrP^{Sc} is done at the Prionics AG as described before (Schaller et al., 1999). The PWB, when validated by the EU with approximately 300 BSE-positive samples from clinically diseased cattle and 1000 negative cattle-brain samples (originating from regions free of BSE), showed a sensitivity and a specificity of 100% (Moynagh and Schimmel, 1999). Western-Blot-negative brain samples are considered as non-diseased and not further investigated. All PWB-positive samples are submitted immediately to the National BSE Reference Laboratory (ITN) for confirmation by histopathology and IHC. Data on the number of cow brains examined with their origin (slaughter plant, rendering plant, or veterinarian) and the results are sent from the Prionics AG to the FVO on a regular basis. Information on PWB-positive test results is immediately forwarded to the emergency slaughter plant, the cantonal veterinarian and the FVO. All detected BSE cases are followed up by questionnaire to collect epidemiological information.

2.5. Data analysis

The databases for clinical BSE suspects, confirmed BSE cases and for all targeted data are maintained in a database management system (MS-Access, Microsoft, Redmond, WA). Descriptive statistics and figures were calculated in MS-Access and in spreadsheet software (MS-Excel, Microsoft, Redmond, WA). Odds ratios and Cornfield 95% confidence intervals or exact confidence limits (whenever appropriate) were derived using Epi Info 6.04 (Centres for Disease Control, Atlanta, GA). For comparison of mean ages at death between groups, a two-sided two-sample t-test assuming equal variances was used.

3. Results

Between 1992 and 1999, on average 81 clinical BSE suspects per year were reported to the veterinary authorities. Between 22 and 69% per year (average 43%) were confirmed as positive cases. The mean annual number of suspects submitted, which proved negative for BSE was 44 (range 31–57) (Table 1).

Collection of fallen stock samples started in January 1999 and the number of samples per month remained relatively stable over time (averaging 562 samples per month). Sampling and testing of emergency-slaughtered cows started at low levels in January and February 1999, but increased to reach the last 12-month average for the period May 1999–April 2000 (386) in May 1999.

The cases detected in the targeted screening resulted in a second peak in the total number of BSE cases by period of reporting (Fig. 1). All targeted-screening cases were born after the ban on feeding of mammalian MBM to ruminants that was implemented in December 1990. Through 30 April 2000, no BSE case has been detected in Switzerland that was born after December 1995 (Fig. 2).

Between 1 May 1999 and 30 April 2000, 104 clinical BSE suspects were reported to the cantonal veterinarians — of which 30 were confirmed with BSE. In addition, 20 BSE cases were detected by targeted screening in the period May 1999–April 2000, 11 in fallen stock and nine in emergency-slaughtered cows. In the period May 1999–April 2000, the odds for finding a BSE case was 49 times higher in the fallen stock than through mandatory

Table 1
Number of cattle suspected to be affected by BSE based on clinical signs of disease, the number of unconfirmed suspects and the number of confirmed BSE cases in Switzerland between 1 January 1992 and 30 April 2000

Category	1992	1993	1994	1995	1996	1997	1998	1999	2000[a]	Average (1992–1999)
Reported BSE suspects	57	68	120	99	95	69	64	77	52	81
Unconfirmed suspects	42	39	57	31	50	31	50	52	39	44
Confirmed BSE cases	15	29	63	68	45	38	14	25	13	37
Proportion confirmed	0.26	0.43	0.53	0.69	0.47	0.55	0.22	0.33	0.25	0.43

[a] Period 1 January 2000–30 April 2000.

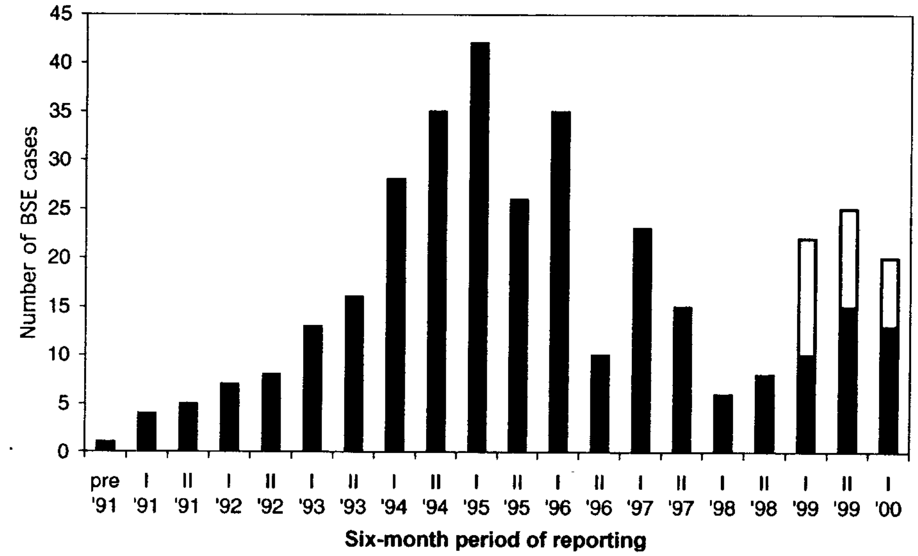

Fig. 1. 349 Swiss BSE cases from passive surveillance (320; black columns) and targeted screening of fallen stock and emergency-slaughtered cows (29; white columns) that were diagnosed through 30 April 2000. Cases are plotted by date of death/reporting over the half-year intervals: 1 January–30 June (I); 1 July –31 December (II).

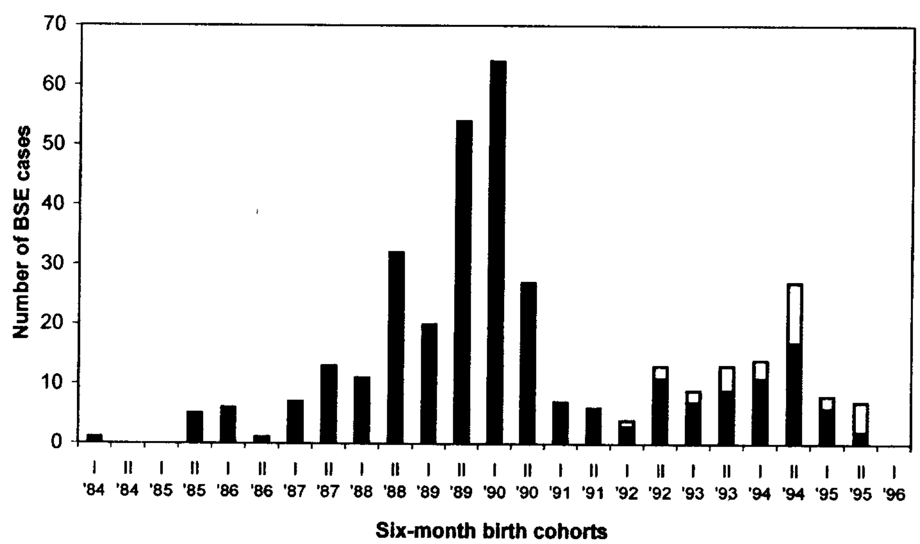

Fig. 2. 349 Swiss BSE cases from passive surveillance (320; black columns) and targeted screening of fallen stock and emergency-slaughtered cows (29; white columns) that were diagnosed through 30 April 2000. Cases are plotted by date of birth over the half-year intervals: 1 January–30 June (I); 1 July–31 December (II). Observation time for the more recent birth cohorts is incomplete.

reporting of clinical suspects from the adult cattle population. For the emergency-slaughter cattle, this OR was 58 and the combined OR was 53 (Table 2).

The mean age at death of all 320 Swiss passive-surveillance cases was 64.5 months (standard deviation, S.D. = 13.9). For the 38 passive-surveillance cases reported between 1 January 1999 and 30 April 2000 this mean was 66.9 months (S.D. = 15.2) — but for the 29 targeted-screening cases detected during the same time period, it was 61.9 months (S.D. = 10.6). The difference (66.9 – 61.9) was not statistically significant ($t = 1.52$, d.f. = 65, $P = 0.13$). The proportion of emergency-slaughter and fallen-stock cases was highest in the age-at-death category 49–60 months (45%). In contrast, for the passive surveillance cases observed during the same time period, this peak occurred at 61–72 months (45%) (Fig. 3).

The number of targeted-screening cases detected was constant over the three 4-month periods May–August 1999 (7), September–December 1999 (7) and January–April 2000 (6), with 4-month incidences of 0.0022, 0.0018 and 0.0018, respectively. The number of passive-surveillance cases initially increased and then decreased during the same time period (5, 16 and 9, respectively). The ratio of passive surveillance to targeted screening cases was 3.2 times higher in the second and 1.5 times higher in the third period when compared with the first period. The differences to the first period were not statistically significant. A similar pattern was observed when the ratios of the respective incidences for the 4-month periods were compared.

All PWB-positive brain samples were confirmed by IHC; however, 80% of the samples from the fallen stock were autolytic and allowed only limited or no pathohistological examination for detection of spongiform changes. Epidemiological trace-back of the

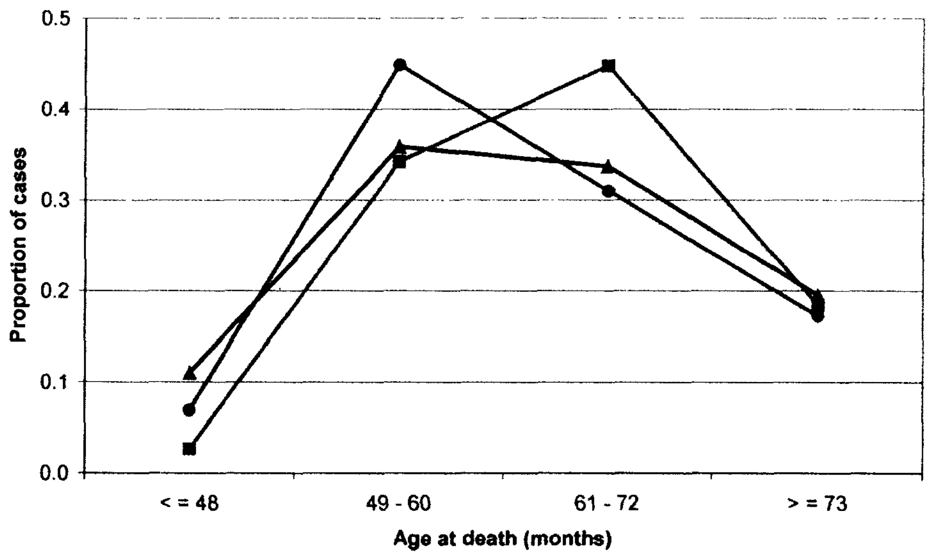

Fig. 3. Proportion of Swiss BSE cases by age-at-death category and method of detection: 282 reported clinical cases through 31 December 1998 (triangles); 38 reported clinical cases since 1 January 1999 (squares); 29 targeted screening cases (emergency slaughter and fallen stock; circles).

Table 2
Results of the Swiss passive BSE surveillance in comparison with the targeted screening of risk populations with the Prionics Western Blot test (active surveillance); reporting period 1 May 1999–30 April 2000

Target population	Total	Cattle tested	BSE cases	12-Month cumulative incidence	Number of cases per 100,000 cows	Odds ratio[a] (95% CI)
Passive surveillance						
Total cow population >24 months	900000	104	30[b]	0.000033	3	Reference category
Targeted screening						
Cows >24 months disposed as fallen stock	6744	6744	11	0.001631	163	49 (23, 102)
Cows >24 months old presented for emergency slaughter	4632	4632	9	0.001943	194	58 (28–128)
Emergency-slaughtered cows and fallen stock combined	11376	11376	20	0.001758	176	53 (29–96)

[a] Odds of detecting a BSE case in the respective target population divided by the odds of capturing a BSE case through passive surveillance; StatCalc module in Epi Info 6.04 (Centres for Disease Control, Atlanta, GA, USA) with Cornfield 95% confidence intervals.

[b] Cases detected through mandatory reporting and subsequent examination of clinically suspect cattle (passive surveillance).

targeted-screening cases revealed that approximately one-third demonstrated clinical symptoms that were typical for BSE, one-third displayed signs of distress, clinical disease and loss of productivity that — in retrospect — should have pointed towards BSE and one-third were removed from the population with clinical signs that cannot be associated with BSE. At the time of culling or emergency slaughter, the owner or treating veterinarian did not classify any of these animals as suspicious for BSE. Details of the follow-up on these cases will be a subject of another study (in preparation).

The total cost of the active targeted BSE surveillance in Switzerland (excluding the personnel at the FVO and the follow-up costs of detecting additional BSE cases) in 1999 accounted for approximately 1 million Euro (close to 1 million US$) (Heim et al., 1999).

4. Discussion

Within a 12-month period 20 BSE-infected cattle close to or actually showing clinical signs of disease that left the population were found by the targeted-screening system in addition to the 30 reported clinical cases detected by mandatory reporting of clinical suspects. In theory, the only other existing exit route for cattle in the Swiss cattle industry is routine slaughter. It cannot be excluded that BSE-incubating cattle without clinical signs are slaughtered. Due to the mandatory ante-mortem slaughter examination, it is considered unlikely (though not impossible) that clearly identifiable clinical BSE-suspect cases will enter this exit route. Nevertheless, to discourage animal owners and veterinarians from using this exit route for suspicious cattle, a random sample of brains from healthy slaughter cows was collected (Doherr et al., 1999b). At present, there is no evidence for the existence of additional exit routes of importance. Also, monthly examination figures for fallen stock and emergency slaughter cows have been relatively constant since May 1999. One major factor that might have biased these results is the difference in age distribution of cattle in the populations targeted for sampling when compared to the general cattle population. The age distributions (and therefore the age-cohort specific incidences of disease) should be taken into account when trying to extrapolate information from these targets to the general population. This is of even greater importance when only a proportion (sample) of the fallen stock and the emergency slaughter targets is examined. At present, however, no reliable age distribution estimates do exist for the fallen stock and the emergency slaughter targets in Switzerland. They should be available after full implementation of the Swiss cattle-registration and movement database that was put in place at the end of 1999. In the meantime, efforts are undertaken to derive these estimates from samples. New models are currently under development within the scope of an European project (FAIRJ-CT98-7021 "Surveillance and Diagnosis of Ruminant TSE") and will take the important information collected in this active-surveillance programme into account when trying to combine the results from different target populations into an estimate for the overall population incidence of detectable BSE cases.

To us, an interesting observation was the higher proportion of targeted-screening cases in the age category 49–60 months (4 years) when compared to the clinical-suspect cases — and the lower mean age of the targeted-screening cases. This could be an indication that the screening test used in this surveillance programme is able to identify BSE-infected cattle several months before they display sufficient clinical signs to be reported as suspect cases (under the current definition of a suspect BSE case). This observation is consistent with results from the pathogenesis studies with experimentally infected cattle done by Wells et al. (1998). The fact that clinical-suspect cases since January 1999 have a higher proportion of cases in the older categories than the clinical cases reported before 1999 could be seen as a first indication for an "ageing" of the remaining BSE cases in the population due to a successful prevention of new infections since 1996. One has to be cautious about drawing final conclusions on these observed age differences until the data are adjusted for the differences in the age structure of the underlying populations.

Slow-spreading diseases with low prevalence and incubation periods close to the productive lifetime of the affected species are difficult to capture in a passive reporting system — even if the collaboration of the owners and veterinarians is assured. This is especially true when decrease in performance is a common early clinical sign because it would often result in culling and replacement of the affected animal without proper diagnosis. When the disease is stigmatised, thought — or officially communicated — not to be present, and confirmation of cases results in the loss of the entire herd, reporting compliance is likely to be low. Improvement of disease awareness (training), changes in disease definition, increase in the compensation offered and other measures might increase this compliance. In contrast, implementation of more stringent disease-control measures, further stigmatisation of the disease and "disease-tiredness" might, over time, reduce case-ascertainment and reporting. Data from scrapie surveillance programs have shown that suspect-case reporting compliance was low (Schreuder et al., 1993; Hoinville et al., 1999; Baumgarten et al., 2000). In this study, the increase in case-ascertainment and reporting by the passive surveillance system seen after August 1999 could be the result of several factors including: (a) renewed awareness for the disease in the context of the implementation of the targeted BSE screening and the publicity going along with it; (b) a change from a whole-herd culling policy (loss of the total herd) through June 1999 to a cohort-culling policy (loss of approximately one-third of the herd) since July 1999; (c) the two continuing-education workshops for large animal practitioners on clinical neurological diseases held in October and December 1999; (d) the difficulty of "hiding" BSE cases because all possible exit routes were covered; (e) the realisation by animal owners and practitioners that BSE-infected cattle — when not reported — might enter the human food chain.

Typical signs are not consistently expressed in animals reaching the clinical phase of BSE and can be confused with those of other diseases (Wilesmith et al., 1992; Hörnlimann and Braun, 1993; Braun et al., 1998a,b, 1999). With the exception of further adaptation of the definition of clinical suspects (for example, by including all cows replaced for decreased productivity such as a drop in milk yield) or by increasing the compensation paid for suspect cases considerably beyond market value, there seems to be little room for improvement of the Swiss passive surveillance system for BSE beyond actual levels.

Using the most recent 12-month period (May–April) instead of the calendar year 1999 ensured that targeted screening of the populations had reached stable levels. This allowed for better comparison of the results over time. The data indicate that the fallen stock and emergency-slaughter groups have a considerably higher proportion of BSE-infected cows than the general population estimates derived from mandatory reporting of clinical suspects would indicate (OR 49 and 58, respectively). BSE-infected animals at the end of their productive (healthy) life seem to accumulate in those cohorts (exit routes) — making these routes good populations for targeted BSE screening (Doherr et al., 1999b). This by no means should discount the value of a passive surveillance system. Before 1999, all detected BSE cases were found among clinical suspects and on average 43% of all BSE suspects in Switzerland were confirmed with BSE. Passive surveillance can be considered as a low-cost screening "test" covering the total population. In the absence of a better ante-mortem screening test for BSE, reporting of suspect cases for further examination is one of the few methods to define risk-populations among living animals.

If all exit routes for detectable (PrP^{Sc}-positive) BSE cases from the population are covered by a targeted screening system, and the surveillance activities are kept constant over an extended period of time, then higher case numbers from the mandatory reporting should lead to lower case numbers from targeted screening (because both approaches are drawing from the same pool of detectable BSE cases in the total cattle population). At the same time, changes in the total number of BSE cases captured in all surveillance activities should reflect true changes in the incidence of detectable BSE cases. In the context of this paper, passive surveillance accounted for 67% of the detected cases over the period of 1 year and targeted screening for 33%. This was accomplished with a high level of disease awareness and the fact that all exit routes from the population were at least partially covered by targeted screening. Also, results from the examination of a routine-slaughter sample have not been included in this assessment (because the number of cases found in the sample was too small to allow for reliable extrapolation). With normal reporting levels and without a targeted surveillance, we believe that it is unlikely that mandatory reporting of clinical suspects will capture >50% of the detectable cases (i.e. adult cattle with sufficient PrP^{Sc} accumulation in the brain and clinical signs of BSE). Conversely, during a recently completed assessment on the geographic BSE risk of EU Member States and Third Countries, it was considered possible that in countries with low prevalence of clinical BSE in the cattle population, this low prevalence has been missed completely by mandatory reporting alone (Anon., 2000a).

For BSE, post-mortem testing for the presence of PrP^{Sc} of sufficiently large samples of fallen stock and emergency-slaughtered cows >24 months old is an important active-surveillance element within a total surveillance system that is based principally on passive surveillance. With the lack of ante-mortem screening tests to detect pre-clinical BSE, a combination of effectively functioning passive and active BSE surveillance strategies as described might be the only approach to assess the BSE situation reliably in a given country or region when the prevalence is low. Disease awareness, stigmatisation and other factors affect passive systems more than active ones. Active-surveillance components are necessary to assess the effectiveness of clinical-suspect reporting and they are considered necessary to substantiate claims of freedom from the disease.

Good estimates for the true BSE incidence are essential to understand the disease epidemiology, required to make sound predictions on the age-specific prevalence of infected animals in the population and on the future course of the epidemic, and important for comparison of the current BSE situations between countries or regions. Within the Office International des Epizooties (OIE) and the European Commission (EC), work is in progress to define requirements for targeted-screening activities within their BSE-surveillance regulations. Both institutions require that risk assessments on the actual risk of introduction and amplification of the BSE agent be done. The Scientific Steering Committee of the EC has concluded that active BSE surveillance results from risk populations such as adult cattle in fallen stock or emergency slaughter could improve the basis for future geographic BSE risk assessment exercises (Anon., 2000a). In the light of the most recently detected first domestic BSE cases in Spain and Germany, the European Commission has decided that the Member States have to implement a targeted-screening of all fallen stock and all emergency-slaughtered cows for BSE as on January 2001. In addition, all must test all healthy slaughter cattle >30 months of age for BSE (Anon., 2000b). The data collected from this targeted BSE screening in all Member States will assist in the further evaluation of the effectiveness of the passive BSE surveillance.

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