Review on the risk for consumption of dairy products produced from sheep's milk obtained from sheep with a positive result for scrapie

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Abstract
Scrapie is a fatal disease that belongs to a group of neurological disorders called transmissible spongiform encephalopathies (TSEs) and affects the brain and central nervous system of sheep and goats. There are two forms of scrapie, classic scrapie and atypical scrapie. There are two forms of scrapie, classic scrapie and atypical scrapie. Classical scrapie can be transmitted from one animal to another, while atypical scrapie is thought to have a spontaneous degenerative disorder in older sheep and is not transmitted to other animals under natural conditions.

Assessing the risk to public health arising from consumption of dairy products produced from sheep's milk obtained from sheep with a positive result for scrapie, we have reviewed the foreign and Bulgarian literary and scientific sources. Based on the analysis, it could be concluded that scrapie is not considered a risk to human health, although it cannot be ruled out with certainty.

Keywords: risk assessment, scrapie, raw milk

Introduction
Transmissible spongiform encephalopathies (TSE), or prion diseases, are fatal neurodegenerative disorders that affect a large spectrum of mammalian species. These conditions include for instance, scrapie in small ruminants, bovine spongiform encephalopathy (BSE) in cattle and chronic wasting disease in wild cervids (CWD). In humans, the most common form of TSE is sporadic Creutzfeldt-Jakob disease (sCJD), which affects ~1–2 individuals per million of the population per year and is generally observed in people aged over 50 years [1].

Scrapie is a fatal disease that belongs to a group of neurological disorders called transmissible spongiform encephalopathies (TSEs) and affects the brain and central nervous system of sheep and goats. This disease is caused by the entry of abnormal prions into the body. Prions are normal proteins in mammals and birds. When an abnormal prion enters a healthy animal organism, it changes the structure of the existing prions and turns them into abnormal infectious prions.

The individual circulating field strains of scrapie in sheep are different pathogenicity, due to which the clinical manifestations of the disease in sheep, goats and mouflons can vary considerably and manifest in different combinations that according to most authors are classified into three main syndromes, including: changes in sensibility, changes in behavior and changes in posture and movements. The clinical signs can last from 1 to 6 months.

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Materials and Methods
With the present work we set ourselves the goal to review the risk arising from consumption of dairy products produced from sheep's milk obtained from sheep with a positive result for scrapie.
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**Results and Discussions**

In the Union food safety legislation, the following acts regulate the monitoring and control on the TSEs, in particular scrapie:

Regulation (EC) No 999/2001 lays down rules for the prevention, control and eradication of transmissible spongiform encephalopathies (TSEs) [7] in animals. It shall apply to the production and placing on the market of live animals and products of animal origin and in certain specific cases to exports thereof. In this Regulation the following are defined as specific risk materials: the skull, together with the brain and eyes, the tonsils, the spine of sheep and goats above twelve months of age or having a permanent incisor that has pierced the gum as well spleen and ileum of sheep and goats of all ages. In case the specifically risky material is not removed from the carcasses of the small ruminants, parts of the body that contain this material or the whole body should be treated as a specific risk material. According to Art. 14 of the Regulation, the Member States shall draw up, in accordance with Community criteria, operational plans and manuals defining national measures to be taken and indicating the competencies and responsibilities in TSE cases. The Bulgarian Food Safety Agency has elaborate guidance with detailed instructions and information, on practical aspects of the fight against scrapie in small ruminant animals [8].


The humans have been naturally exposed to the agents that cause scrapie in small ruminants for at least 200 years, but despite large-scale epidemiological studies, no direct evidence of transmission of scrapie to humans has been found. Much evidence and data have been published in the scientific literature on the potential for transmission of the causative agent of classical scrapie through milk. The European Food Safety Authority (EFSA) has reviewed the results of these studies in its scientific opinions on the risks to human and animal health. These opinions contribute to the understanding of scrapie transmission mechanisms as a prion-induced disease.

The 2007 EFSA opinion [2] on certain aspects related to the risk of TSEs in ovine and caprine animals concluded that there was no evidence of an epidemiological or molecular link between classical and/or atypical scrapie and TSEs in humans.

In 2008, EFSA published another opinion [3] on the dangers of TSEs to humans and animals from exposure to milk and dairy products from small ruminants, concluding that classical scrapie could be transmitted from ewes to lambs through milk or colostrum and that the use of milk and dairy products from a herd with classical scrapie may carry a risk of TSE exposure for humans and animals. With regard to atypical scrapie, EFSA points out that the limited (according to observations) spread of its causative agent in the body of the affected animals could limit its transmission through milk.

The main finding of the last EFSA opinion of 2015 [4] confirms the conclusion made in 2011 European Center for Prevention and Control joint scientific report [5] that there is no scientific evidence that classical scrapie can be transmitted from sheep to humans in natural / real conditions. In this regard, a study was reviewed that showed that under laboratory conditions, classical scrapie can be transmitted from sheep to mice whose genes have been modified to mimic human genes. This transmission caused the appearance of a disease similar to the sporadic form of Creutzfeldt-Jakob disease (sCJD). These models have limitations in extrapolating the results to natural conditions, both in terms of how well they represent the species barrier in humans and how well the mode of experimental contamination reproduces exposure under natural conditions. On this basis, it is considered that although the risk of transmission of TSE agents to humans in ovine animals cannot be ruled out, this risk is extremely low.

According to EFSA, the biodiversity of pathogens in ovine and caprine animals is a key element that does not preclude the possibility of human transmission, and that it is this diversity that increases the likelihood of any of these agents being transmissible. However, EFSA acknowledges that there is no scientific evidence to show a direct link between TSEs in sheep and goats other than bovine spongiform encephalopathy and TSEs in humans.

In November 2008, the French Food Safety Authority (AFSSA) published an opinion on the risk of classical scrapie spreading in the milk of small ruminants, drawing the same conclusion as that of EFSA with regard to the possibility of transmitting classical scrapie. Scrapie from the ewe lamb through milk or colostrum [6]. With regard to TSE exposure for humans, AFSSA is of the opinion that the consumption of milk and milk products from infected herds of small ruminants or from herds suspected of being infected should not be allowed, as it could lead to excessive consumer exposure.

Regarding atypical scrapie in sheep and goats, the available scientific data are too limited to conclude whether this disease has zoonotic potential.

**Conclusions**

**Based on the above, it could be concluded that**

Scrapie is not considered a risk to human health, although it cannot be ruled out with certainty. Although the risk of human transmission of TSE agents in ovine and caprine animals cannot be ruled out, this risk is extremely low as evidence that interspecific transmission is possible, based on the experimental models that do not reproduce the natural conditions associated with the true species barrier in humans and the actual modes of infection.

Given the established case of classical scrapie, the combination of low levels of infectivity in milk and low prevalence of the disease, combined with the lack of direct data on a definite link between the classical scrapie and the transmissible spongiform encephalopathies in humans, as well as and the use of a mixture with a predominant amount of milk from healthy animals suggests that the consumption of sheep cheese produced under ban will expose human health to a very low risk of contracting scrapie.

Given that it is an established case of atypical scrapie, there is no risk to human health from the consumption of the said cheese, as the spread of the causative agent of this disease in
the body of diseased animals is limited and has not yet been identified in peripheral tissues, such as the mammary glands, which limits its transmission with milk.

References
2. EFSA. Opinion of the Scientific Panel on Biological Hazards on certain aspects related to the risk of Transmissible Spongiform Encephalopathies (TSEs) in ovine and caprine animals. 8 March. The EFSA Journal. 2007; 466:1-10.