# Transmission and characterisation of Swiss cases of atypical small ruminant TSE in ovine transgenic mice

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**Key words**
Transmissible spongiform encephalopathies, prion, small ruminants, BSE, scrapie, atypical scrapie, ovine prion protein transgenic mice

**Aim of the study**
The pathobiology of atypical scrapie, a prion disease in sheep and goats, is still poorly understood. In a preceding study (Nentwig et al, 2007), we demonstrated that atypical scrapie affected small ruminants differ in the neuroanatomical distribution of the pathological prion protein ($PrP^d$). The aim of this study was to investigate whether these differences depend on host- or rather pathogen- related factors.

**Material and methods**
We transmitted scrapie from Swiss atypical scrapie isolates to ovine prion protein transgenic mice and compared survival times, attack rates, patterns of lesions, $PrP^d$ distribution in the brain and biochemical $PrP^d$ characteristics between these isolates and with those of classical scrapie, BSE and a Nor98 control.

**Results and significance**
Our results demonstrate that atypical scrapie in Switzerland, despite phenotypic variations in the natural hosts, is caused by a uniform type of prion clearly distinct from those involved in BSE and classical scrapie. The observed phenotypic differences in small ruminants are, therefore, likely host-dependant. Our results also point to a previously unrecognized complexity of the molecular $PrP^d$ phenotype. These findings show similarities to other prion diseases in animals and humans and lay the groundwork for future comparative research.

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