



## Stable tubule only polypeptides (STOP) proteins co-aggregate with spheroid neurofilaments in amyotrophic lateral sclerosis

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Résumé en anglais	<p>A major cytopathological hallmark of amyotrophic lateral sclerosis (ALS) is the presence of axonal spheroids containing abnormally accumulated neurofilaments. The mechanism of their formation, their contribution to the disease, and the possibility of other co-aggregated components are still enigmatic. Here we analyze the composition of such lesions with special reference to stable tubule only polypeptide (STOP), a protein responsible for microtubule cold stabilization. In normal human brain and spinal cord, the distribution of STOP proteins is uniform between the cytoplasm and neurites of neurons. However, all the neurofilament-rich spheroids present in the tissues of affected patients are intensely labeled with 3 different anti-STOP antibodies. Moreover, when neurofilaments and microtubules are isolated from spinal cord and brain, STOP proteins are systematically co-purified with neurofilaments. By SDS-PAGE analysis, no alteration of the migration profile of STOP proteins is observed in pathological samples. Other microtubular proteins, like tubulin or kinesin, are inconstantly present in spheroids, suggesting that a microtubule destabilizing process may be involved in the pathogenesis of ALS. These results indicate that the selective co-aggregation of neurofilament and STOP proteins represent a new cytopathological marker for spheroids.</p>
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## Liens

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