

Results: 20 B-MB were identified; by WHO definition, most of these resided within the classic category (N = 19), while one was LCA. 13 of 20 B-MB displayed ‘scattered’ nodules; by molecular subtype, these included eight group 4, four group 3 and one WNT tumors. Seven of the 20 B-MB exhibited ‘frequent’ nodules; by molecular subtype, these included six group 4 and one group 3 tumors. Statistical analysis confirmed this non random distribution of B-MB across molecular subtypes.

Conclusion: Our data confirm the work of Ellison et al. that suggested B-MB is genetically different than DN-MB. In particular, B-MB resides in the non-WNT/SHH molecular category, but especially amongst group 4 when nodularity is ‘frequent’.

5. Automated analysis of 1p/19q status by FISH in oligodendroglial tumours.

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Automated analysis of 1p and 19q status in oligodendroglial tumors by fluorescence in-situ hybridization (FISH) can be achieved by image-analysis software present in the majority of institutions using the FISH technique. Despite the widespread availability of this software, there are no specific guidelines in the literature on how to use it.

We studied which green/red (G/R) probe signal combinations are predictive of 1p/19q co-deletion in a retrospective series of 53 oligodendroglial tumours and defined a new algorithm with a reduced sequence of combinations compared to previous studies. This algorithm was then tested and refined on a prospective series of 45 oligodendroglial tumours. The new algorithm scores 24 G/R combinations, which represent less than 50 % of the total observed combinations in our series. This algorithm excludes some previously described combinations and redefines the place of others. G/R combinations of 5/2, 6/2 and 6/3 associate with deletion status combinations, combinations of 1/2 associate with normal chromosome status, and combinations of 3/3 and 4/4 associate with imbalanced chromosome status.

The new algorithm when applied to the combination and ratio methods of signal probe analysis gives a high concordance between manual and automated analysis on examination of 100 tumour cells (91% concordance for 1p and 89% concordance for 19q) and total concordance on examination of 200 tumour cells. This highlights the value of automated analysis to identify cases with imbalanced chromosome status, in which a larger number of tumour cells should be studied by manual analysis. Our algorithm can be easily programmed on all existing FISH analysis software platforms and should facilitate multicentric evaluation and standardization of 1p/19q assessment in gliomas.

6. Surfen, a proteoglycan antagonist, reduces lysolecithin-induced demyelination with related effects on macrophage function

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Proteoglycans are components of the extracellular matrix and have roles in brain development and responses to injury. Connective tissue components are known to be major inhibitors of remyelination in mouse models of demyelination and are found at the border of active demyelinating lesions in Multiple Sclerosis. Surfen (bis 2-methyl, 4- amino, 6-quinolyl amide) is a small molecule antagonist previously shown to bind preferentially to heparan sulfate and related proteoglycans.

We have previously reported that surfen reduces T cell proliferation *in vivo* and *in vitro*. Here we report the effects of surfen on an *in vivo* model of demyelination and its effects on macrophage function *in vitro*. Demyelination was induced by injecting the detergent lysolecithin into the spinal cord dorsal columns of adult C57Bl/6 mice. Relative to vehicle treated mice, co-injection of surfen (100 µM) with lysolecithin reduced total lesion area seven days post-injection. Because macrophages dominate these lesions and influence remyelination, murine bone marrow derived macrophages were assessed using assays of chemotaxis and phagocytosis. Macrophages chemotaxis was increased in response to surfen (10 µM) relative to vehicle by approximately 15% (p < 0.05). Phagocytosis of *E. coli* was not affected by surfen.

These effects of surfen on experimental demyelination and macrophage function suggest that proteoglycan binding may promote aspects of myelin repair relevant to Multiple Sclerosis.

7. The pathogenesis of Friedreich cardiomyopathy: myocarditis

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Fra-taxin deficiency causes the complex neurological and cardiac phenotype of Friedreich ataxia (FRDA). The most common cause of death is cardiomyopathy. The results presented here are based on a systematic study of fixed and frozen archival heart specimens and include measurement of cardiomyocyte hypertrophy, fra-taxin assay, X-ray fluorescence (XRF) of iron (Fe) and zinc (Zn), inductively-coupled plasma optical emission spectrometry of these metals in digests of left ventricular wall (LVW), right ventricular wall (RVW), and ventricular septum (VS), Fe histochemistry, and immunohistochemistry and double-label immunofluorescence microscopy of cytosolic and mitochondrial ferritins, and of the inflammatory markers CD68