



Articles appearing in the May 2020 issue

Improvement of stiff-person syndrome symptoms in pregnancy case series and literature review

Objective To describe 2 cases from a single academic institution of improvement in stiff-person syndrome (SPS) symptoms during pregnancy and to review the clinical outcomes of SPS in 6 additional pregnancies described in the literature.

Methods Evaluation of clinical symptoms and treatment changes of disease state during pregnancy.

Results Seven patients with 9 pregnancies are described in women with a diagnosis of SPS. Six of 7 (86%) women were positive for glutamic acid decarboxylase (GAD65) antibody. In 5 of 9 (56%) pregnancies, symptomatic medications (antispasmodics) were significantly reduced with stabilization or improvement in symptoms through pregnancy. Nine live, healthy pregnancies resulted. All 7 (100%) women experienced worsening of symptoms after the birth of their children, and symptomatic therapies were resumed and/or increased.

Conclusions The immune pathogenesis of SPS continues to be explored. Immunomodulatory shifts during pregnancy may influence changes of clinical SPS symptoms and provide insight into the unique pathogenesis of SPS. Some women with SPS may be able to reduce symptomatic medications related to clinical improvement during pregnancy. Women with SPS may safely carry pregnancies to term, delivering healthy and unaffected babies.

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Cortical topological network changes after optic neuritis

Objective To differentiate between visual cortical network topology changes after optic neuritis (ON) stemming from different inflammatory disease types, we used mathematical graph theory—based tools to analyze functional imaging data.

Methods Sixty-two patients were recruited into this cross-sectional study, 23 of whom had neuromyelitis optica spectrum disorder (NMOSD) with ON, 18 with clinically isolated syndrome (CIS)-ON, and 21 with other CIS episodes. Twenty-six healthy controls (HCs) were also recruited. All participants underwent resting-state functional MRI. Visual networks were defined using 50 visual regions of interest. Analysis included graph theory metrics, including degree, density, modularity, and local and global efficiencies.

Results Visual network density shows decreased connectivity in all patient groups compared with controls. A higher degree of connections is seen in both ON groups (CIS and NMOSD) compared with the non-ON group. This pattern is most pronounced in the dorsal-lateral regions. Information transfer efficiency and modularity were reduced in both CIS groups, but not in the NMOSD group, compared with the HC group.

Conclusions Visual network density seem affected by the neurologic deficit sustained (ON), and connectivity changes are more evident in the dorsal-lateral regions. Efficiency and modularity seem to be associated with the specific disease type (CIS vs NMOSD). Thus, topological cortical changes in the visual system are associated with the type of neurologic deficit within the limits set on them by the underlying pathophysiology. We suggest that cortical patterns of activity should be considered in the outcome of the patients despite the localized nature of ON.

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