Infantile spasm syndrome

Infantile spasm (IS) syndrome is an age-related epileptic encephalopathy that occurs in children.

ReHo values of left precuneus and right superior frontal gyrus were associated with the epilepsy duration in IS. The correlation results indicate that the involvement of these regions may be related to the seizure generation. The results suggest that IS may have an injurious effect on the brain activation. The findings may shed new light on the understanding the neural mechanism of IS epilepsy ¹⁾.

Diagnosis

MRI is very helpful to locate the pathology in the pediatric population, since only a small portion had a normal MRI ²⁾.

Outcome

Spasms of focal onset have a similar postsurgical outcome as other seizure types so surgery may be an excellent option for treating selected patients with focal infantile spasms. Volume and type but not topography of the lesion influence the age of onset $^{3)}$.

Case series

2017

Chipaux et al. retrospectively reviewed all children with IS referred to ther tertiary center between 2002 and 2014 and try to define factors of outcome.

Sixty-eight children with focal onset seizures were referred: twenty children with a hemispheric implication and 48 with one or more lobes involved. The age of onset was significantly earlier in the hemispheric population (8.0 versus 16.7 months in the focal population). There was no difference in the age of onset between anterior and posterior onset zones, as we could expect regarding the maturation gradient. The epilepsy began earlier in life in tuberous sclerosis than in DNET. Only three children of the 48 non-hemispheric patients had a normal MRI at the time of the surgery. Temporal lobe was involved only in a third of the population. More than 86% of the patients were operated on. Patients with hemispheric lesions were operated on younger (2.6 years+/- 2.1 years) compared to 4.6+/- 3.5 years in the whole population. The most frequent etiologies were in descending order: dysplasia, ganglioglioma or dysembryoplastic tumours and tuberous sclerosis. The global seizure outcome was favorable (Engel 1a) in 74.6% of the patients, and 87.9% if the delay between the first seizure and the surgery was less than 36 months. It fell to 64.7% if the delay exceeded 50 months.

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2016

The purpose of a study was to investigate regional homogeneity (ReHo) changes in IS patients. Resting state functional magnetic resonance imaging was performed on 11 patients with infantile spasm syndrome, along with 35 age- and sex-matched healthy subjects. Group comparisons between the two groups demonstrate that the pattern of regional synchronization synchronization in IS patients is changed. Decreased ReHo values were found in default mode network, bilateral motorrelated areas and left occipital gyrus of the patient group. Increased ReHo was found in regions of cingulum, cerebellum, supplementary motor area and brain deep nucleus, such as hippocampus, caudate, thalamus and insula. The significant differences might indicate that epileptic action have some injurious effects on the motor, executive and cognitive related regions. In addition, ReHo values of left precuneus and right superior frontal gyrus were associated with the epilepsy duration in the IS group. The correlation results indicate that the involvement of these regions may be related to the seizure generation. The results suggest that IS may have an injurious effect on the brain activation. The findings may shed new light on the understanding the neural mechanism of IS epilepsy ⁵⁾.

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