

Role of botulotoxin type a in the complex treatment of the hypokinesia in patients with Wilson disease

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Background: Hypokinesia is common extrapyramidal sign in patients with Wilson disease; it leads to significant disturbances of life quality and reduces independence of personal living skills. Aim: to evaluate effectiveness of Botulotoxinum in complex treatment of rigidity and extrapyramidal signs in female patient with trembling and rigidity symptoms. Methods and materials: female patient (onset of disease – 8y.). She was on permanent complete bed rest due to severe hypokinesia and rigidity; no voluntary movements were possible; severe joint contractures; permanent hyperkinetic disorders and severe pseudobulbar syndrome were described. Patient was on constant therapy (dopaminergic, neurotrophic, copper eliminating agents). Results: due to non-sufficient results of constant base line therapy Botulotoxinum (Dysport®, Bofur Ipsen, 1000 U) was added. Positive result was observed starting from the 14th day. It has been resulted in appearance of voluntary movements in upper and lower limbs, increase of movements, and decrease of hyperkinesia and pain syndrome. Evaluating of injected muscles showed significant decrease of muscle hypertonia during 1 month (from grade 4 to 2 Ashworth scale). After 1, 5 month after injection patient managed to eat by herself; after 3 months managed to change position from bed to wheel chair without assistance. Conclusion: Complex therapy of severe Wilson disease using Botulotoxinum was recognized as effective.