

## Two steps individualized ACTH therapy for West syndrome

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**Objective:** To minimize adverse effects of ACTH therapy in good responders and to obtain maximum treatment effects in poor responders, we introduced a new therapeutic regimen of two steps individualized ACTH therapy for West syndrome (WS).<sup>1,2</sup>

**Methods:** This is a prospective multi-center study, carried out between 1996 December and 2000 January. Twenty infants with newly diagnosed WS, cryptogenic (4), symptomatic (16), who had failed to respond to high-dose vitamin B6 (40-50mg/kg/d) and zonisamide (10-12mg/kg/d), were enrolled into the two steps individualized ACTH therapy study. Synthetic corticotropin (ACTH-Z 0.025mg/kg/dose, max 0.25mg) was first administered intramuscularly every other day seven times in 14 days. The poor responders with persisting epileptic spasms received additional ACTH therapy every day for 1-2 weeks followed by tapering of the drug. Seizure and EEG outcome, i.e., disappearance of epileptic spasms, hypsarrhythmia and adverse effects were looked for up to 1 month after completion of ACTH.

**Results:** After the 1st step of ACTH therapy, epileptic spasms disappeared in 10/20 infants (50%), cryptogenic WS in 2/4 infants (50%), symptomatic WS in 8/16 infants (50%). Hypsarrhythmia disappeared in 10/17 infants (59%), cryptogenic WS in 2/4 infants (50%), symptomatic WS in 8/13 infants (62%). Rare and mild adverse effects only were observed. Nine out of 10 infants with persisting epileptic spasms received the 2nd step additional ACTH therapy. Epileptic spasms disappeared in 3/9 infants (33%), cryptogenic WS in 1/2 infants (50%), symptomatic WS in 2/7 infants (29%). Hypsarrhythmia disappeared in 3/7 infants (43%), cryptogenic WS in 1/2 infants (50%), symptomatic WS in 2/5 infants (40%). There were more adverse effects compared to after the first step. Combining both steps of ACTH therapy, epileptic spasms disappeared in 13/20 infants (65%), cryptogenic WS in 3/4 infants (75%), symptomatic WS in 10/16 infants (63%). Hypsarrhythmia disappeared in 13/20 infants (76%), cryptogenic WS in 3/4 infants (75%), symptomatic WS in 10/13 infants (77%).

**Conclusion:** The efficacy of two steps individualized ACTH therapy was similar to previous reports of conventional ACTH therapy. Individualization of the ACTH therapy according to treatment response resulted in shorter treatment duration and less adverse effects in good responders, and maximum treatment effects in poor responders.

### References

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