Brain compensatory mechanisms enhance the recovery in the Rasmussen encephalitis child patient: a case was followed for 16 years

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To the Editor: Rasmussen encephalitis (RE) is a chronic inflammatory disease of unknown origin, which usually affects one hemisphere of the brain.1 It is an uncommon disease of the nervous system that is usually seen in children. While the cause of RE is unknown, current evidence suggests that autoimmune processes are responsible for the damage to the brain cells. Because of repeated convulsions and progressive neuronal damage, patients often suffer from mental decline and neurological impairment.

A 12-year-old girl presented to the hospital with a paroxysmal left limb twitching and exacerbation of left limb weakness. She was apparently well until October 2004 when she had the first episode. Later on, she gradually developed paroxysmal involuntary tremors in the left lower extremity, a few seconds each time, 2 to 3 times per day. She has consciousness, no headache, nausea, vomiting, binocular gaze, fatigue, and so on. In December 2004, the patient suddenly lost consciousness during the class and fell to the ground with limb tetanic convulsions, binocular gaze, no incontinence, and bite of tongue. After 3 min, the symptoms of seizures disappeared. Since then, the girl also developed recurrent seizures. The magnetic resonance imaging (MRI) of the cranial and spinal cord was normal [Figure 1A]. The electroencephalogram (EEG) showed slow waves on the top of the right occipital lobe. She was diagnosed as viral encephalitis and secondary epilepsy. After treatment, the frequency of the above attacks was dramatically reduced, and the amplitude of limb shaking was decreased. However, the frequency of seizures suddenly increased in September 2005, with more than ten attacks per day. The girl had disturbance of consciousness, incontinence, headache, vomiting, and weakness in the left limb.

The girl was born in Guiyang, Guizhou Province. The patient went on vocation in February 2004 before episode. There is no history of hepatitis and tuberculosis vaccination, and no pets in the family.

The score was 30 points on the mini-mental state examination (MMSE) scale. Limb muscle strength is level 5 and muscle tension is normal. The tendon reflex of left upper and lower limb (+ + + +), it includes the radial nerve of the brachioradialis muscle, biceps musculocutaneous nerve, triceps radial nerve, and quadriceps (patellar) femoral nerve. Left patellar clonus and ankle clonus (+). The tendon reflex of right upper and lower limb (+ +), Babinski sign (−), Pussep sign (+) on the left. Kernig sign (−) in October 2004.

When the girl was presented to hospital for the second time, a review of the cranial MRI revealed the cortex and subcortical irregular lamellae on the right parietal occipital lobe were slightly longer T1, longer T2 signals, the fluid attenuated inversion recovery (FLAIR) phase was high, the boundaries were clear, and the diffusion-weighted imaging (DWI) and diffusion tensor imaging were not diffuse. Restricted, the lesion near the sulcus narrowed slightly, and abnormal signals increased, and no abnormalities in conventional, biochemical, or TOACH results of cerebrospinal fluid (CSF). The number of seizures was reduced after treatment, but the weakness of the left limb was not
improved. The right parietal occipital lobe of the cranial MRI was enlarged compared to previous one. Her rehabilitation included learning, writing, using a computer, and walking every day.

There were no abnormalities in blood routine, routine urine, blood biochemistry, and coagulation. Hepatitis B, human immunodeficiency virus (HIV), syphilis, and hepatitis C antibodies were negative. Erythrocyte sedimentation rate, serum ammonia, and anti-streptococcus hemolysin O antibody are normal. Thyroid function, copper and serum copper levels and serum ceruloplasmin were normal. The chest radiograph was normal. Lumbar puncture (2005-9-12): CSF pressure 150 mmH2O, CSF biochemistry, routine and rubella virus, herpes simplex virus, cytomegalovirus, toxoplasma IgM, IgG antibodies were all normal. HIV antibody (-). Immunological examination (2005-9-12) showed that rheumatoid factor (-), antinuclear antibody (-), antineutrophil cytoplasmic antibody (-), extractable nucleur antigen antibody profile (-), normal immunoglobulins (IgA, IgG, IgM), C-reactive protein is normal. Bilateral brainstem auditory evoked potential (2005-9-13) showed no abnormalities. No seizures were observed by Video EEG (2005-1-25), and sharp slow waves were detected during awake and sleep stages in the right parietal occipital lobe. Muscle biopsy (2005-10-10) suggested that muscle fibers had increased lipid droplets and some type II muscle fibers atrophy. Brain stereotactic biopsy (2005-10-11) indicated neuronal degeneration in the right cortex, mild loss, a large number of heterotypic cells reactive hyperplasia, infiltration of CD8 positive microglia and a small number of lymphocytes, scattered nerve cells. Electromyography (2005-09-28) results suggested that no obvious neurogenic and myogenic damage was found in the left and upper limb, and no obvious abnormalities were found in the left brachial plexus and bilateral lower limb sciatic nerve branches.

During the latest follow-up on August 28th, 2019, the patient was currently engaged in copywriting and her educated level was high school. She was in a good mental state and could walk by herself. She had dragged on her left lower extremity, occasional small involuntary left forearm jitter. Physical examination showed that the speech was fluent and left facial was paralyzed. Muscle tension of left limb was higher than the right. Muscle strength of left upper limb was 4-level and that of left lower limb grade 4. Pathological signs were not elicited. The MMSE score was 30 points. The cranial MRI (2019-11-22) confirmed the right cerebral hemisphere was extensively softened (long T1, long T2 signal, low signal on FLAIR), glia hyperplasia and perforation deformity (cavitated brain). The NAA peak of the residual brain parenchyma significantly decreased, the Cho peak and Cr peak did not change much, and no diffusion-restricted area was seen on DWI. The white matter fiber bundles in the affected side of the brain were significantly reduced, and most of the interruptions were absent. Cho: Choline; Cr: Creatine; DWI: Diffusion-weighted imaging; FLAIR: Fluid attenuated inversion recovery; MRI: Magnetic resonance imaging; NAA: N-acetylaspartate.

**Figure 1:** (A) The cranial MRI (2004-12-08). There was no obvious abnormality. (B) The cranial MRI (2019-01-01). The right cerebral hemisphere was extensively softened (long T1, long T2 signal, low signal on FLAIR), glia hyperplasia and perforation deformity (cavitated brain). The NAA peak of the residual brain parenchyma significantly decreased, the Cho peak and Cr peak did not change much, and no diffusion-restricted area was seen on DWI. (C) The cranial DIT (2019-12-11). The white matter fiber bundles in the affected side of the brain were significantly reduced, and most of the interruptions were absent. Cho: Choline; Cr: Creatine; DWI: Diffusion-weighted imaging; FLAIR: Fluid attenuated inversion recovery; MRI: Magnetic resonance imaging; NAA: N-acetylaspartate.
children’s brains are more plastic. Previous study also found that the neural network connections in the remaining side of the brain of these people were stronger.\[3\] This study demonstrates that people with only one hemisphere of the brain not only showed complete normal neural network connections, but also had a similar connectivity pattern compared to those people with two hemisphere brain. Insights from these rare patients argue that intrinsic mechanisms of brain organization in only half of the typically available cortex can be sufficient to support extensive cognitive compensation.\[4\] Language function has also shown near-complete recovery in many patients who had their language-dominant hemisphere resected.\[5\] Here, we proposed that some circuits can perform similar functions. Another explanation is that the patient’s hemisphere brain developed alternative neural circuits for performing the specific functions. The cranial MRI suggests that the neural networks can compensate for each together as long as those important neural networks are not destroyed. All in all, our study suggested that the circuit and function of the brain can be reshaped by the training. We will continue to follow up and do related genetic testing.

**Acknowledgements**

The authors thank the patient.

**Conflicts of interest**

None.

**References**


**How to cite this article:** Long HC, Liu CF, Chu L. Brain compensatory mechanisms enhance the recovery in the Rasmussen encephalitis child patient: a case was followed for 16 years. Chin Med J 2020;133:2880–2882. doi: 10.1097/CM9.0000000000001083