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Paroxysmal episodes, "re-build up" phenomenon and moyamoya disease

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ABSTRACT – Moyamoya disease is an idiopathic cerebral vasculopathy, which may be progressive or non-progressive. Non-idiopathic forms with an associated disease are called moyamoya-like syndrome. The electroencephalographic finding characteristically seen after hyperventilation in about 50% of children with cerebrovascular disease includes gradual frequency decrease and activation of amplitude of slow waves which appear after the disappearance or attenuation of ordinary build up. This is termed the *"re-build up" phenomenon*, which is rarely seen and therefore may be under-recognized. We present video telemetry during a transient ischaemic event of a child subsequently diagnosed with moyamoya-like syndrome. We highlight the potential for misdiagnosis of organic non-epileptic events. Hyperventilation during EEG should be avoided in children with known moyamoya disease. *[Published with video sequences]*

Key words: paroxysmal episodes, re-build up, moyamoya disease, EEG hyperventilation, transient ischemic event

Case Report

We report a case of a 6 year old girl who had a normal delivery at term. From 18 months she had episodes termed "funny turns" by her parents. EEG at four years showed slow wave disturbance over the left posterior quadrant with focal epileptiform activity. Neurometabolic investigations were normal. Brain MRI showed right parietotemporal atrophy. She was diagnosed as having epilepsy and started on sodium valproate and subsequently topiramate.

At 5 years of age concerns emerged regarding deterioration in school performance and co-ordination difficulties. Because of concerns regarding clinical history and discordant EEG and MRI findings she had video telemetry which captured a habitual attack precipitated by her crying for 15 minutes *(figure 1A, B)*, with marked truncal

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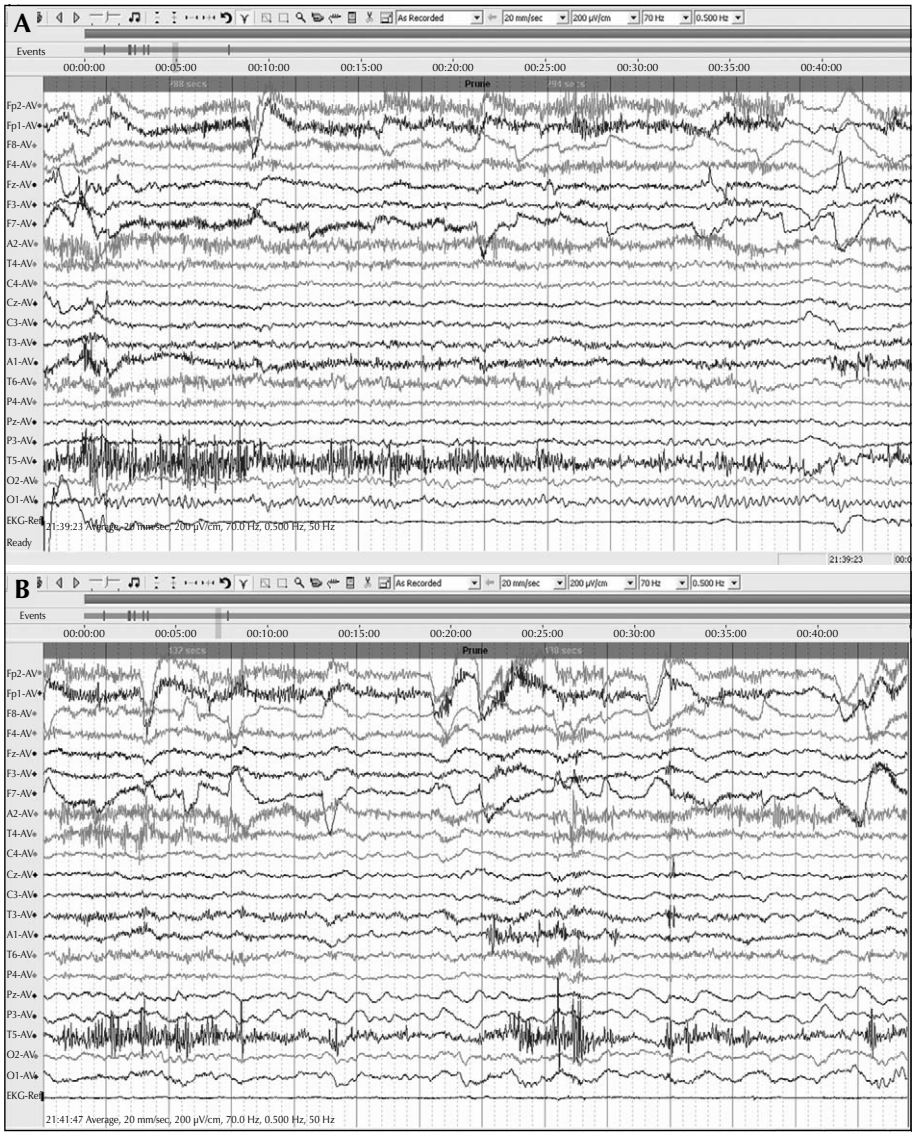

Figure 1. A) Child starts crying as father is about to leave. The EEG is contaminated by artefact, however normal posterior alpha rhythm is seen bilaterally. **B**) The child is seated on her mother's lap and still crying. Note EEG change with appearance of focal slowing over the left parieto-occipital region.

ataxia and right sided weakness lasting three minutes *(figure 2, see video sequence)* accompanied by slowing over the left parietal region. Her EEG then normalised as she settled *(figure 3)*.

Review of previous EEGs *(figure 4)* showed appearance of generalised delta after the end of hyperventilation lasting for up to three minutes consistent with "re-build up", and an habitual episode of becoming floppy after hyperventilation *(figure 5)*.

In one of the previous recordings she had been noted to be floppy after hyperventilation.

Telemetry and EEGs were suggestive of transient focal ischaemia in the left parietotemporal region of the left middle cerebral artery.

Review of MRI scans *(figure 6)* showed multiple bilateral infarcts and flow voids within the basal ganglia secondary to parenchymal collaterals.

Moyamoya disease was confirmed on cerebral angiography *(figure 7)*. Investigations for stroke risk factors were normal. The child had a left ECIC by-pass, with a reduction in transient ischaemic symptoms. She remains on anti-platelet and anti-epileptic medication.

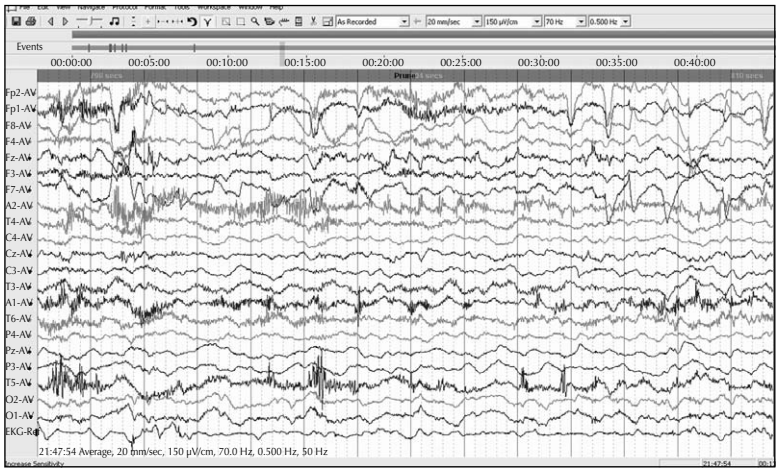

Figure 2. Event-note marked truncal ataxia and right sided arm weakness as the child stands up. Note EEG slowing over left posterior quadrant.

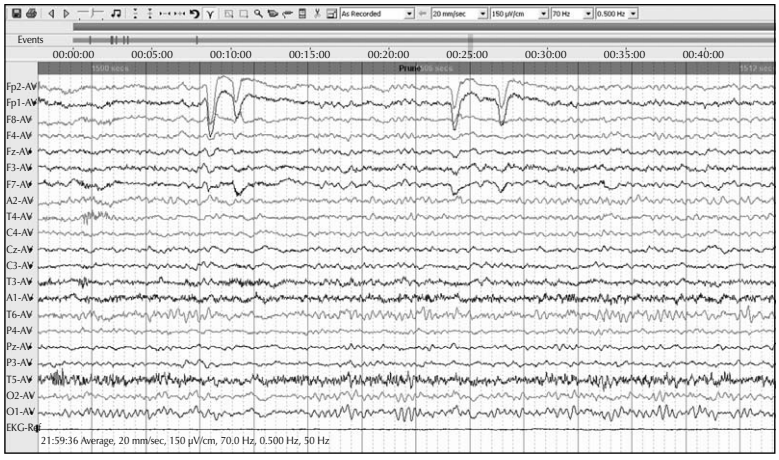

Figure 3. Post event-EEG has normalised.

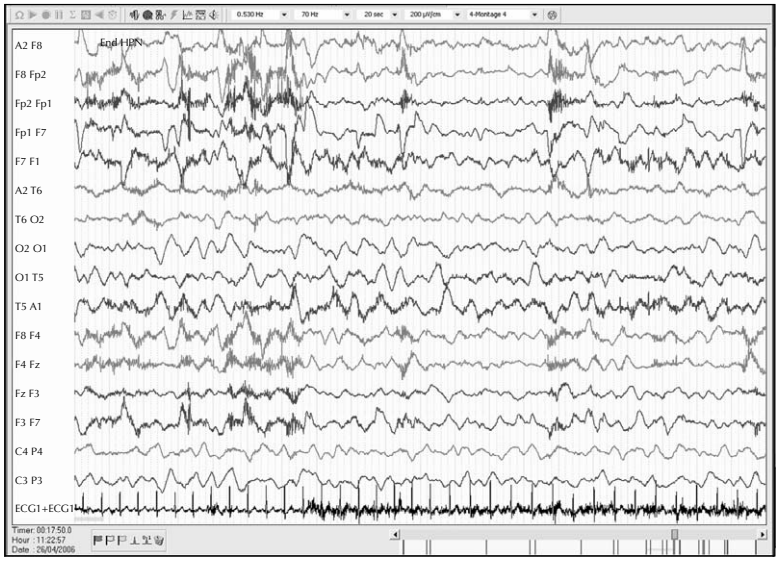

Figure 4. EEG shows background slowing associated with hyperventilation, which attenuates once the child stops over breathing.

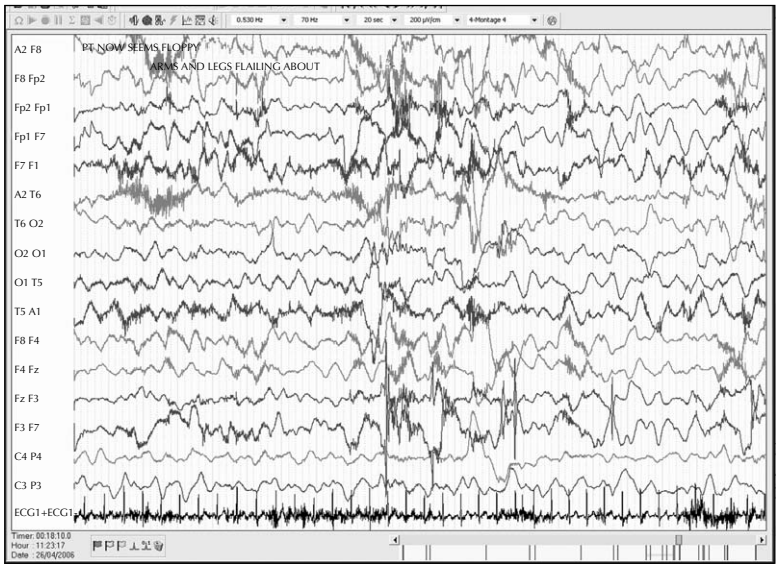

Figure 5. EEG shows re-build up of diffuse slow waves 30 seconds after the child has stopped hyperventilating, most marked over the bifrontal and left posterior quadrant. The child is noted to be symptomatic.

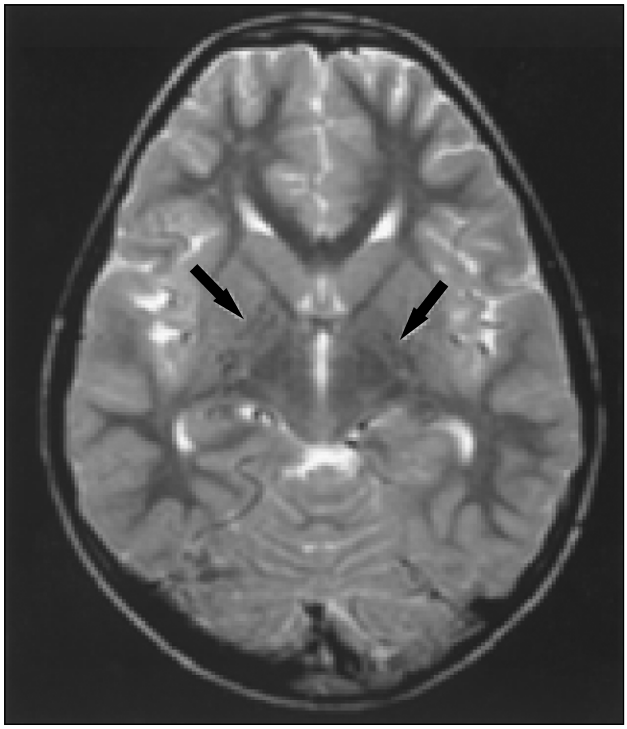

Figure 6. T2 weighted MRI showing multiple flow voids (arrows) within the basal ganglia and bilateral frontal areas of cortical infarction and atrophy.

Conclusion

"Re-build-up" may be an unrecognized EEG finding of moyamoya disease. It represents the uncoupling between metabolic demand and blood supply to the cortex (Kuroda *et al.*, 1995, 1996) and cerebrovascular insufficiency. This case highlights the potential for misdiagnosis of organic non-epileptic events and the importance of video telemetry and multidisciplinary review of complex cases. Hyperventilation during EEG should be avoided in children already diagnosed with moyamoya disease. □

Disclosure.

None of the authors has any conflict of interest to disclose.

Legend for video sequence

Video telemetry showing habitual paroxysmal episode. Note that she has marked truncal ataxia, is floppy with poor head control, right sided weakness and dystonic movements particularly noticeable as she attempts to stand and hold the bed, accompanied by EEG slowing in the left parietal region.

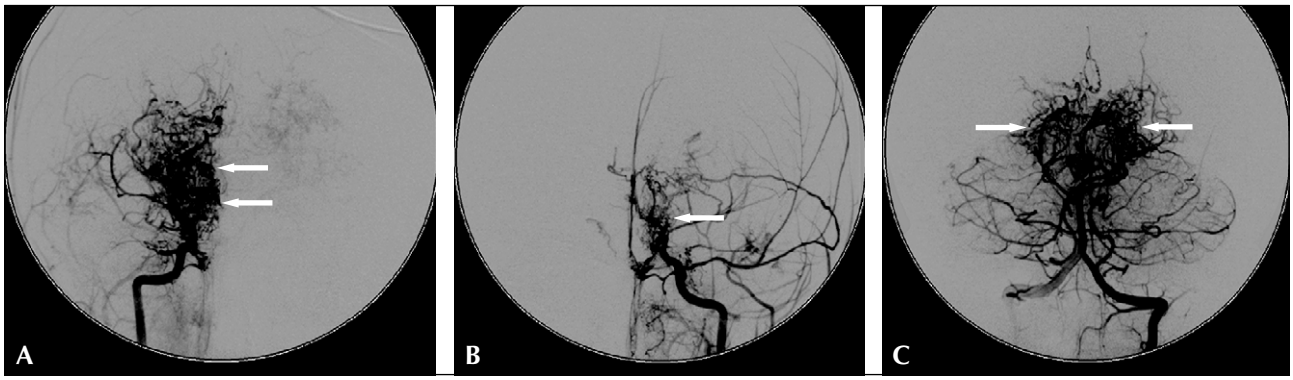

Figure 7. Cerebral angiography of the right (**A**) and left (**B**) internal carotid and left vertebral (**C**) arteries showing multiple areas of stenoses involving the terminal internal carotid arteries. Additionally, there is narrowing of the anterior, middle and posterior cerebral arteries with extensive collateralization at the base of the brain, referred to as "puff of smoke appearance" (arrows), seen in moyamoya disease.

References

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