CASE 3

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A baby boy of 19 months, who had a mild upper respiratory infection, choked on a piece of candy, vomited repeatedly for 15 minutes and became deeply unconscious. He recovered consciousness shortly after admission and was discharged from hospital four days later with no residual symptoms.

Three months later he was readmitted semiconscious with twitching of the right side of his face and right arm and leg. Both pupils were widely dilated and reacted sluggishly to light and accommodation. He appeared under-developed and his mother stated he could not walk, sit or crawl. Cerebrospinal fluid was clear and of normal pressure. He gradually became unconscious, though his right sided twitchings were no longer apparent. He died two months later without regaining consciousness.

There were five children in this family, born in the following order:

(1) A girl, born in 1941, who is alive and well. The present father is her step-father.
(2) A boy, born in 1951, who developed an upper respiratory infection, pneumonia and generalized convulsions, and died at 14 months of age.
(3) A boy, born in 1952, who developed choreiform twitchings in all limbs and died one month later aged 24 months.
(4) A boy, born in 1953, who is alive and well.
(5) The present case, a boy, born in 1960.

On gross inspection of the brain there was slight thickening of the leptomeninges in the interpeduncular fossa and slight symmetrical dilatation of the lateral and third ventricles.