Narcolepsy was recently found to be strongly associated with HLA-antigens. All patients with narcolepsy showed 100% positive HLA-DR2 through studies from Japan, U.K., France, and Canada. It was suggested that the presence of HLA-DR2 was a sine qua non for the development of narcolepsy, not vice versa, though, those having HLA-DR2 could not always become narcoleptic. Some triggering factors would be necessary for the manifestation of narcolepsy, though it is not clear at present whether these factors may be genetic and/or environmental. To elucidate the problems concerned, it is much effective to observe MZ twins incompletely or completely discordant for narcolepsy. We have experienced two pairs of MZ twins completely discordant for narcolepsy.

A male patient with narcolepsy, who was born in 1924 as the first-born of MZ twins. Five of the six siblings were healthy. A brother of his mother was suffering from excessive daytime sleepiness (EDS). Genetic markers were identical. Blood groups: O, MNss, CCDee, Fy(a+b-), Jk(a+b-), P1. Ear wax: dry. PTC(+), MDH(-), Hp(2-1). HLA: A2, A9, B15, Cw1, Cw3. DR2 of the patient was positive, but that of the co-twin’s was not yet identified. The probability of being MZ was 0.983. The patient started to sleep frequently at his office after the age of 30, and was given a nickname, “Nemuri-Kyoshiro”, denoting a sleepy fellow. At the age of about 40, cataplexy on emotional excitement, hypnagogic hallucinations and sleep paralysis were developed. His birth weight was 1700g, which was smaller than his twin brother’s. He was also inferior to his twin brother with regard to manual dexterity and physique during their elementary school days. After graduating from junior high school, the patient became a resident employee of a bookstore and attended an evening business school for 5 years. His twin brother worked at an electrical company and never tried to get advanced education. The patient suffered from hilus tuberculosis at the age of 16. He was later employed at a government office, and attended an evening college for another 6 years. His twin brother did not experience EDS, and rather was a short-time sleeper, and had some difficulty in falling asleep.

Another female patient, born in 1942, was the second-born of MZ twins. The first-born’s delivery was entirely normal in contrast to that of the patient, who had a breach presentation and asphyxia neonatorum. Sleep attack, cataplexy and hypnagogic hallucinations were appeared at the age of 12. The probability of being MZ is 0.9700.