

HSAN IV affects males and females in equal numbers. Several hundred cases have been reported in the medical literature. The exact incidence and prevalence is unknown. More cases have been reported in Japan than any other country and the frequency of the disorder is higher in the Japanese and Israeli-Bedouin populations due to a founder effect. A founder effect is when a small isolated population of settlers (founders) expands over several generations leading to a high prevalence of a particular genetic trait. Onset of the disorder is at birth.