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Clinical features in the VHL disease in Japan: A survey for the onset of age on the VHL associated diseases.

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Information for VHL disease, including the numbers of family, incidences and characteristics of VHL associated tumors is not clarified yet in Japan. To make more accurate information for this disease, we have performed survey studies for several times. We analyzed the onset age in the VHL disease from a survey study in the total number of 180 families that are supposed to consist of almost of the VHL families in Japan here. The mean onset age in CNS hemangioblastoma(HB), retinal angioma(RA), renal cell carcinoma(RCC) and pheochromocytoma(pheo) is 27.9, 25.9, 34.5, or 30.2 years old, respectively. Onset of each disease has broad peak and minor small peak. The onset of age in RCC is earlier than that in European countries. The onset of age in RA and pheo is older than that in European countries. Since the types of germline mutation in Japan is not so much difference in the onset age is due to the difference. This information makes presymptomatic clinical diagnosis much easier and promote better prognosis for this disease in Japan.

Key words: onset age, RCC, RA