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1: Am J Kidney Dis. 2001 Oct;38(4):770-6.

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Am J Kidney Dis

Familial clustering of ruptured intracranial aneurysms in autosomal dominant polycystic kidney disease.

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Ruptured intracranial aneurysm (RICA) is a life-threatening complication of autosomal dominant polycystic kidney disease (ADPKD). A family history of RICA may be a risk factor for RICA. Six hundred eight adult members of 199 ADPKD families were interviewed, and family pedigrees were constructed. Individuals were classified as having definite, probable, or possible RICAs from evidence and history obtained in interviews. Central nervous system (CNS) events not consistent with RICA were classified as other CNS events. Seventy-seven CNS events occurred in 906 subjects with ADPKD (8.5%) versus 13 events in 823 subjects without ADPKD (1.6%; $P < 0.0001$). No event in subjects without ADPKD was consistent with an RICA. Twenty-seven other (non-RICA) CNS events occurred in subjects with ADPKD (3%) versus 13 events in subjects without ADPKD (1.6%; $P = 0.05$). The frequency of RICA was increased in subjects with ADPKD: 21 definite RICAs in subjects with ADPKD (2%) versus none in subjects without ADPKD ($P < 0.001$); 28 definite and probable RICAs in subjects with ADPKD (3%) versus none in subjects without ADPKD ($P < 0.001$); and 50 definite, probable, and possible RICAs in subjects with ADPKD (5.5%) versus none in subjects without ADPKD ($P < 0.001$). The null hypothesis that RICAs are randomly distributed among subjects with ADPKD was tested for definite RICAs ($n = 21$), definite and probable RICAs ($n = 28$), and definite, probable, and possible RICAs ($n = 50$). In the three categories, the null hypothesis was rejected at P less than 0.05, P less than 0.05, and P less than 0.005, respectively. Vascular CNS events occurred more frequently in ADPKD than non-ADPKD family members, and clustering of RICAs occurred in families with ADPKD.

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