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Recurrence of intracranial aneurysms in autosomal-dominant polycystic kidney disease

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Recurrence of intracranial aneurysms in autosomal-dominant polycystic kidney disease.

Background. The natural history of intracranial aneurysms (ICAs) in individuals with autosomal-dominant polycystic kidney disease (ADPKD) is poorly defined.

Methods. We followed twenty ADPKD subjects, eleven with ruptured and nine with intact ICA, for 15.2 ± 8.1 years (range, 6.0 to 33.2years). Initial diagnosis was by four-vessel cerebral angiography in eighteen subjects. Follow-up examinations were four-vessel cerebral angiography in fourteen and magnetic resonance angiography (MRA) in six subjects. We examined the occurrence of new ICAs, an increase in size of existing ICAs, recurrent rupture or surgical intervention, and death.

Results. Age at initial diagnosis of ICA was 37.7 ± 10.4 years (range, 20.2 to 53.1 years). Seventeen subjects (85%) had an anterior and three (15%) had a posterior ICA at initial diagnosis. On restudy, five subjects (25%) had a significant change, consisting of new ICAs in a different location in all five and an increase in size of an existing ICA in two of the five. All subjects with ruptured ICA and one subject with intact ICA had undergone surgery at the time of initial diagnosis. Ten subjects (50%) underwent further surgery 8.1 ± 6.1 years later (1.3 to 17 years). No subject died during follow-up and one subject experienced a recurrent RICA (RICA). We were unable to identify risk factors associated with development of a new ICA or increase in size of an existing ICA.

Conclusion. Individuals with ADPKD and ICA appear to be at moderate risk for new ICAs and increase in size of existing ICAs; mortality and risk of recurrent rupture, however, appear to be low.

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