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1: [Am J Gastroenterol.](#) 2005 Nov;100(11):2569-82.



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### New advances in evaluation and management of patients with polycystic liver disease.

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Adult polycystic liver disease (APLD) is an autosomal dominant condition most commonly associated with polycystic kidney disease. However, over the last decade it has come to be recognized that APLD is a genetically heterogeneous disorder involving derangements on at least three different chromosomes. Mutations involving chromosomes 16 and 4 accounting for autosomal dominant polycystic kidney disease (ADPKD) type 1 and type 2 have been well described as have their gene products, polycystin-1 and polycystin-2. These have since been joined by a more recently recognized mutation in the short arm of chromosome 19 thought to be responsible for a much rarer form of autosomal dominant polycystic liver disease without any associated renal involvement. Despite the sometimes impressive physical and radiologic findings, only a minority of patients will progress to advanced liver disease or develop complications as a result of massive hepatomegaly. In these patients, medical management alone has proved ineffectual. Therefore, in the symptomatic APLD patient, surgical therapy remains the mainstay of therapy and includes cyst aspiration and sclerosis, fenestration with and without hepatic resection and orthotopic liver transplantation. The surgical literature on treatment of APLD, to include outcome measurements and complication rates are summarized. Additionally, we review other potential organ involvement and resultant complications.

PMID: 16279915 [PubMed - indexed for MEDLINE]

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