Title: Autosomal Recessive Polycystic Kidney Disease – *PKHD1 GeneReview* Hepatobiliary Pathology

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Hepatobiliary Pathology (Biopsy or Autopsy)

The histologic findings of developmental ductal plate anomalies, including bile duct proliferation, biliary ectasia, and periportal fibrosis, are present in all individuals with ARPKD [Kamath & Piccoli 2003, Jiang et al 2016].

- The hepatobiliary disease is the result of a developmental defect in which a failure of ductal plate remodeling results in persistence of embryologic bile duct structures that eventually can become massively dilated.
- Associated portal veins are often abnormal, demonstrating dilatations and an increased number of smaller portal vein branches.
- Significant fibrosis may be seen in the portal tract even at birth, and as affected children age, the amount of periportal fibrosis increases, resulting in hepatomegaly and progressive portal hypertension.

Interestingly and for unclear reasons, livers often demonstrate proportionally larger left lobes than right lobes [Gunay-Aygun et al 2013].

References

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