Vascular Anomalies in Alagille Syndrome

To the Editor:

We read with interest the article by Kamath et al about vascular anomalies in Alagille syndrome (AGS). We know that hypotheses are not anyone’s property, but it is unfair to omit two previously published papers from our team. Indeed, we suggested after studying JAGGED1 expression during human embryogenesis that abnormal angiogenesis was implicated in the pathogenesis of AGS and particularly the paucity of interlobular bile duct. Furthermore, we recently suggested that defects of Notch signaling pathway may impair both angiogenesis and hemostasis in AGS patients and that arterial endothelial cells, which mainly express JAGGED1, play a pivotal role.

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Response

We appreciate Dr Hadchouel’s comments about our article. Our study was designed as a clinical review of vascular anomalies in a large cohort of patients with Alagille syndrome. We did not intend our article to be a comprehensive or exhaustive review of the literature supporting the hypothesis that a vasculopathy underlies Alagille syndrome. We recognize the enormous contribution of Dr Hadchouel and her group to the study of Alagille syndrome and in particular their studies relevant to a vasculopathy in this condition. A review of all the relevant literature was beyond the scope of the discussion in our paper, but we are grateful to all the groups studying Alagille syndrome who help to develop our collective hypotheses.

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