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LETTER TO THE EDITOR

Frequency and pathogenesis of central liver nodules in Alagille syndrome patients: Reply to Libbrecht and Cassiman

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Dear Editor,

We appreciate Libbrecht and Cassiman [1] for taking interest in our article titled "Giant hepatic regenerative nodules in Alagille patients" by Rapp et al. [2] and for providing insight into the etiology of these giant hepatic nodules. Libbrecht et al. [3], both in their current letter to the editor and in prior publications, have discussed a plausible explanation for the development of these giant hepatic regenerative nodules in Alagille patients.

The aims of the paper by Rapp et al. were to depict and discuss the imaging features of these nodules; among the limited publications, ours was a large series. Unlike Libbrecht et al., the focus of our work was not on the nodules' pathological etiology. This could be considered a limitation of our manuscript. In our paper, we note that the etiology of these giant regenerative nodules was not well established, yet we did not explicitly state that the etiology was unclear. We acknowledge the previous work by Libbrecht and colleagues where they have made clear the defect in the postnatal bile duct branching and elongation lies at the heart of the development of these nodules.

We also agree that it would be helpful to know at what time point these giant regenerative nodules start to develop in Alagille patients. Our study was retrospective and spanned a 10-year period. In all our cases, we identified the giant regenerative nodule years after a child was diagnosed with Alagille, often after the development of cirrhosis and portal hypertension. We also agree with Libbrecht and colleagues regarding the notion that children who develop giant regenerative nodules are unique; neither all our Alagille patients nor those in prior publication by Alhammad et al. [4] developed these nodules. Perhaps performing periodic serial imaging studies earlier in the course of the disease in this Alagille population will provide clues to the etiology, development and natural course of these giant regenerative nodules.

Compliance with ethical standards

Conflicts of interest None.

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