AJNR

Reply:

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AJNR Am J Neuroradiol 2011, 32 (7) E143
doi: https://doi.org/10.3174/ajnr.A2650
http://www.ajnr.org/content/32/7/E143

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We thank Dr McKinney and colleagues for their interest in our article. The authors generally agree with our findings and raise several interesting issues. We are grateful to the Editor-in-Chief for the opportunity to respond to some of the main issues raised.

Dr McKinney writes that we described only lesions in the cingulate and insular cortex. This is not the case. Throughout our article, we stated that all our cases had additional findings such as more variable additional cortical involvement or basal ganglia changes.[1] The point we were rather hoping to put across was that while such changes were not specific (eg, diffuse cortical involvement could be seen in a range of conditions including diffuse hypoxic injury), symmetric involvement of the cingulate and insular cortex was seen in all 4 cases and could therefore represent a more specific finding for hyperammonemic encephalopathy. Such symmetric findings are not commonly seen in other conditions, as far as we are aware.

Dr McKinney and colleagues also argue that the terms "acute hyperammonemic encephalopathy" and "acute hepatic encephalopathy" could be interchangeable. They then suggest that their proposed terminology would be more accurate, given that in acute hepatic failure, hyperammonemia is only 1 of the precursors of encephalopathy.[2] Respectfully, we have to disagree. The use of this terminology would have to assume that all cases of hyperammonemic encephalopathy are caused by hepatic failure; this is also not the case. In our small series, 3 patients had hepatic failure, but 1 patient was post–orthoptic lung transplant without evidence of hepatic failure. Moreover, in the pediatric literature, there are many described cases of hyperammonemic encephalopathy, without any link to liver failure.[3,4] Many of these cases, (eg, due to urea cycle disorders) had radiologic findings similar to ours with cingulate and insular cortex involvement.[4] We argue that this provides compelling evidence that the changes we describe are related to hyperammonemic encephalopathy rather than hepatic encephalopathy per se. We believe that these findings can be seen in hyperammonemic encephalopathy due to various causes, such as hepatic failure, urea cycle disorders, post–lung transplantation, and so forth.

It is unclear from our data whether cortical involvement lies at the severe end of the spectrum, as Dr Mc Kinney suggests because 2 of our patients survived despite having cortical involvement. This survival suggests that these changes could potentially be reversible. Data from other series hopefully could shed more light on prognosis. Only 1 of our patients had brain stem and thalamic involvement, and we are unable to comment further on the basis of our small series.

In summary, we suggest that the pattern of symmetric cingulate and insular cortical involvement, with additional and more variable cortical involvement, should alert the radiologist to the possibility of hyperammonemic encephalopathy. The cause of the hyperammonemia is nonspecific and could include hepatic failure, lung transplantation, urea cycle disorders, and so forth. Despite the differences in viewpoints, both Dr Mc Kinney's and our articles should increase the radiologic recognition of this potentially reversible condition.[1,2]

References

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DOI 10.3174/ajnr.A2650