Analysis and Classification of Cerebellar Malformations

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To the Editor: We were very interested in the article "Analysis and Classification of Cerebellar Malformations" by Patel and Barkovich (1), and we thank the authors for their imaging-based classification of cerebellar malformations, which highlights an area of great confusion. This classification has an important value because it defines and helps in the recognition of some cerebellar malformations that have been less described in the radiologic literature.

We ask ourselves whether an imaging-based classification represents categories of similar malformations. Even if it is very helpful to understand these malformations, it may be artificial to separate a first category of "cerebellar hypoplasia" from "cerebellar dysplasias" because most of the malformations included in the latter category are associated with vermian hypoplasia. Moreover, to quote prenatal "cytomegalovirus" infection as an example, its etiopathogenesis and prognosis are probably not the same as the prognosis of "isolated diffusely abnormal foliation." We think that a classification based on cerebellar embryology, genetics, or signaling molecules that play a role in specifying cerebellar domains (morphogenesis) but also in cellular migration (histogenesis) (1, 2) would better define different types of cerebellar malformations that are similar in terms of their consequences to brain function. Nevertheless, we agree that to obtain such classification is a very difficult task.

We were also very interested in and satisfied with the description and the recognition of focal and/or diffuse cerebellar cortical dysplasia as a new radiologically detectable cerebellar malformation. Although the pathology literature has already provided relevant information about its morphologic characteristics, it has recently been revealed by MR imaging and deserves note in the radiology literature. We agree that focal cortical dysplasia will be recognized more commonly, especially in patients with developmental disabilities. Even if the mechanism by which the dysplasia develops remains poorly understood, its recognition could probably be the first step to understanding its role.

The authors report that imaging findings in cases of cortical dysplasia and cerebellar heterotopia have not been previously described and that, moreover, they do not have histologic details of isolated focal cerebellar dysplasias. We bring to their attention our previous report of cerebellar cortical dysplasias already published in the *AJNR* (3) and our article concerning the neuropathologic description of a neonatal case of isolated focal cortical dysplasia associated with cerebellar heterotopia (4). Even if the pathogenesis of focal cerebellar cortical dysplasias is being investigated (4), its pathologic significance and the mechanism by which the same malformation that is often associated with more widespread brain anomalies could be, in other cases, an isolated finding remains unknown.

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Reply

We thank Dr. Soto-Ares for his kind comments regarding our article (1) and extend our compliments on his very interesting article in the same issue of the *AJNR* (2). We agree with his comments that a classification based on embryology, genetics, or signaling molecules is optimal for the description of malformations; indeed, this subject was discussed in our article. However, as Dr. Soto-Ares acknowledges, such a classification is very difficult and we are a long way from having enough knowledge of cerebellar genetics and molecular biology to accomplish such a classification. For the present, we think that a radiologic classification is a useful first step in studying this complicated group of diseases, just as it has proved useful in studying cerebral cortical malformations (3, 4) and anomalies of the cerebral commissures (5, 6).

Regarding Dr. Soto-Ares' question about the pathogenesis of focal cerebellar cortical dysplasias in the absence of cerebral anomalies, we can suggest several possibilities, including focal ischemia or infection, somatic mosaicism, or different dosages of the same mutation in the same gene in different parts of the CNS. We agree that these are important questions that are not possible to answer by radiologic investigation.

We are delighted to have interested and enlightened colleagues such as Dr. Soto-Ares working in this field of study. The contributions of Dr. Soto-Ares and his colleagues to the study of cerebellar malformations have been both interesting and enlightening (2, 7). As advances in this field result from the cooperation of all those interested in the subject, we are happy that he has found our article worthy of comment and we hope for further interactions with his group and with others who have interest in cerebellar development and maldevelopment.

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