

## Origins of the 5mm Chiari Rule

If you're a Chiari veteran, you've probably heard it over and over...Chiari is 5mm or more of tonsillar herniation. But do you know where this definition came from? It didn't come from Hans Chiari, the pathologist who first described several cases (which incidentally were not representative of what we think of as Chiari I today), and it didn't come from Arnold, whose students somehow got his name attached to the disorder for a period of time. In fact, like far too many things that are taken as given in science and medicine, the origins of the 5mm rule are a bit murky and sadly do not have a solid scientific basis. In the 1980s, the advent of MRIs provided an opportunity to study Chiari malformation non-invasively and also quantitatively. While it is likely not the whole story, a publication in 1986 (Barkovich) by a group of radiologists used MRI to measure the tonsillar position and is often cited as the origin of the 5mm rule. The study involved 25 Chiari cases and 200 healthy controls. The Chiari group was selected as having definite Chiari clinical signs and symptoms. The MRIs by today's standards were low powered and low resolution. The researchers used hard copies of the images for their measurements. They did find that in the control group, the tonsillar position ranged from 8mm above the foramen magnum to 5mm below and there was overlap between the groups in the 3-5mm range. The researchers then calculated the accuracy of using different cut-offs between 1-5mm. From this data, over time people interpreted that Chiari should be at least 3-5mm of herniation. For reasons that are not clear, this later evolved to 5mm. Interestingly, the authors in the Barkovich paper stress in their discussion of the results that it is better to err on the side of inclusion and to not rule out Chiari too early, and therefore suggest a cut-off of 2mm. Of course, the problems and limitations with these cut-offs in general have been discussed extensively in past research updates and indeed were laid bare by Milhorat in the late 90s with his seminal article showing that symptomatic Chiari does exist with smaller herniations. More recently, studies by Meadows and Maher have shown that 1-3% of the general population may meet the 5mm rule for Chiari, but that only a small fraction of these will ever be symptomatic. As the survey discussed above shows, the Chiari community is now at the point where experts discount the 5mm rule and researchers are looking aggressively for a new way to define Chiari malformation.

**Source:** *Significance of cerebellar tonsillar position on MR. Barkovich AJ, Wippold FJ, Sherman JL, Citrin CM. AJNR Am J Neuroradiol. 1986 Sep-Oct;7(5):795-9.*