

Chiari & Epilepsy In Children

For some time, physicians and researchers have been aware of a subgroup of pediatric Chiari patients that also have epilepsy (or vice versa). When these cases first started to be reported in the medical literature, there was speculation that the two conditions were linked in some way. For example, was some aspect of the cerebellar tonsils being herniated or disruption of the flow of spinal fluid causing epilepsy? Did both Chiari and epilepsy arise from some other, unidentified issue? Or did both conditions share a common genetic source? Limited research since then has failed to find a causative association or a common underlying issue.

So on the other hand, it is also possible that the associations seen in these patients is not linked, but due to chance. Seizures are fairly common, with some estimating that as many as 10% of people will experience at least one seizure in their lifetime. It is also estimated that epilepsy affects between .6-1% of children, and because it involves in the brain anyone with a seizure will undergo imaging. Research has shown that as many as 3% of all children meet the 'MRI' definition of Chiari with 5mm or more of tonsillar descent, so invariably some of the children with epilepsy will also have Chiari on MRI.

In an attempt to shine some light on the subject, a team of Italian doctors compared two groups of pediatric patients. The first group was comprised of 29 children who were initially seen during a 5 year period for epilepsy and then later found to have tonsillar herniation of 5mm or more on MRI (Chiari). The second group was comprised of 77 pediatric Chiari patients seen during that same time frame who also underwent EEG testing (which can show brain wave abnormalities).

In Group 1, 27% had Chiari specific symptoms (in addition to epilepsy) and 31% underwent decompression surgery. All of the epilepsy cases were treated with medication. At the end of the study, 62% of the group were seizure free. In Group 2, 75% underwent decompression surgery and while 17% had EEG anomalies, none had epilepsy.

When the team compared the two groups, they found no significant differences in the male/female ratio, percent of cases with syringomyelia, tonsillar improvement after surgery, or symptom improvement after surgery. The only significant differences between the groups were that Group 1 was younger and Group 2 had larger herniations. Further, in Group 1 there as no correlation between Chiari surgery and the seizures resolving. Similarly, in Group 2 there was not connection between Chiari surgery and EEG anomalies resolving.

Based on this, the authors conclude that clinically, epilepsy and Chiari can be considered separate conditions and treated accordingly; meaning medication for epilepsy and surgery for Chiari when warranted. While they failed to find any evidence that links the two, they do point out that Group 1 represented 12% of the epilepsy cases and 18% of the Chiari cases seen overall at their institution during that time. This is higher than you would expect by chance, but the authors also point out that their institute may attract this specific type of case.

However, the Chiari1000 also shows higher numbers than you would expect by chance with 7% of the pediatric participants (175) reporting also being diagnosed with epilepsy and 15% with other types of seizures.

But beyond these numbers, for now no evidence has been found of a connection between the two.

Source: Chiari 1 Malformation and Epilepsy in Children: A Missing Relationship. Massimi L, Palombi D, Contaldo I, Veredice C, Chieffo DRP, Calandrelli R, Tamburrini G, Battaglia DI. J Clin Med. 2022 Oct 20;11(20):6182.

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