

Consensus Statement on Pediatric Abnormal Craniocervical Motion with Chiari

Craniocervical instability, or abnormal craniocervical motion (ACCM), in the presence of Chiari is more commonly found among adults than children. Data from the Chiari1000 indicates that roughly 10% of adults report also being diagnosed with instability compared to only 2.8% of children. Because it is uncommon in children, it is not routinely screened for. Like adult Chiari related instability, it also lacks a precise definition, and there can be variations among doctors on when to look for it, how to look for it, and when to say it exists.

Recently, a group of 14 pediatric neurosurgeons issued a consensus statement on the topic of ACCM in pediatric Chiari in order to provide guidance on when to screen for it, how to screen for it, and how to define it. They chose the term ACCM as opposed to instability because they felt that the term instability implied incorrectly that surgical stabilization is required in all cases. The international group of surgeons were highly experienced with extensive Chiari practices. Eighty percent had more than 15 years of experience and 60% had performed more than 200 Chiari decompressions during their careers. Highlighting how infrequent the issue of ACCM is among children, 90% of the surgeons estimated they perform five or fewer craniocervical fusions per year.

The process to develop the consensus statement involved the group responding to an initial survey about their clinical practice and beliefs. The responses were then used to form draft statements which the individuals then agreed or disagreed with. Statements with at least 80% agreement were considered to have reached consensus. A meeting was held to discuss statements which were close to consensus and the final document was drafted. Each surgeon agreed to the publication of the final document.

The consensus statement included 22 separate points: 8 focused on when to screen for ACCM, 3 on the type of imaging to use, 7 on the specific measurements and symptoms that are indicative of ACCM, and 4 on the use of a hard collar to further diagnose ACCM.

The group agreed that in general pediatric Chiari patients without bony anomalies or genetic conditions should NOT be screened for ACCM unless they met one of three criteria:

- 1. Assimilation of the atlas, where the top vertebra (the atlas) is congenitally fused to the bottom of the skull
- 2. Recurrent classic Chiari headaches or bulbar symptoms after decompression surgery. Bulbar symptoms refer to symptoms that arise from dysfunction of the lower brainstem and involve the muscles of the face, tongue, and throat which lead to problems with speech, swallowing, and chewing
- 3. Unchanged or increasing syringomyelia with or without associated symptoms after decompression surgery

The group also agreed that the imaging of choice should be upright x-rays and that use of CTs and MRIs is not recommended. The group identified a number of imaging measurements that should be looked at and recommended specific cut-off values for ACCM. The details of these measurements are beyond the scope of this article. Finally, the group agreed that in patients with imaging suggestive of ACCM that a trial immobilization with a hard collar can be used to further the diagnostic process. If symptoms improve with a hard collar, and imaging supports it, then the patient should be considered to have ACCM.

It is not clear what impact a consensus statement like this will have on clinical practices beyond those who participated in the process, but at a minimum it provides parents with an educational tool when ACCM is suspected.

Source: Alexiades NG, Anderson DI, D'Ambrosio A, et al. Building consensus regarding the definition of abnormal craniocervical motion in pediatric patients with Chiari malformation: a modified Delphi study. J Neurosurg Pediatr. Published online March 7, 2025. doi:10.3171/2024.11.PEDS24478



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Conquer Chiari is a 501(c)(3) public charity dedicated to improving the experiences and outcomes of Chiari patients through education, awareness and research.