

Study Tracked Natural History of Syringomyelia for 10 years+

In recent years there have been a growing number of Chiari related publications coming out of China. Now, a group there has published the results of a study which tracked the natural history of Chiari related syringomyelia for more than ten years. Specifically, the study looked at 156 adult CM/SM patients who were initially treated conservatively – meaning no surgery – and followed with clinical visits and imaging for at least ten years to identify characteristics which predicted both deterioration and spontaneous resolution.

The doctors based their initial treatment decision in large part on the Japanese Orthopedic Association (JOA) score, which is widely used in Asia to assess damage to the cervical spine. Patients with scores of 13 or above (out of 17) were considered for conservative management. Obvious deterioration was then defined as the JOA score dropping at least 2 points to below 13. Spontaneous resolution of the syrinx was defined as a 50% or more reduction in the length, width, or both on imaging. Patients with obvious deterioration underwent surgery and were subsequently removed from the study.

In terms of final outcomes, the group was split roughly into thirds, with 33% experiencing spontaneous resolution, 35% obvious deterioration, and 32% remaining stable. It is important to note that the resolution rate is only out of patients who were considered mild to start and the resolution rate of all CM/SM patients evaluated during the study time period was 4.3%. The doctors found a number of differences between patients who deteriorated and those who didn't, such as syrinx location, syrinx size, a specific neurological reflex, and the presence of sleep apnea at diagnosis. However, when they looked at all the potential variable together, they found that the presence of sleep apnea and a positive Babinski sign were the most predictive of eventual deterioration.

The Babinski test involves stroking the sole of the foot firmly with something like a pen. In adults, if the big toe curls upwards and the other toes fan out, it is considered a positive response and indicative of damage to the nerves in the spine. Interestingly, in babies this response is considered normal. For the purposes of this study, sleep apnea was defined as an average of 5 events or more per hour as assessed by a sleep study.

The team performed a similar analysis to identify characteristics that predicted spontaneous resolution. They found that patients whose syrinxes shrank on their own were more likely to have syrinxes located only in the cervical region and to NOT have sleep apnea.

This study is a great example of the type of data that can be invaluable to patients (and clinicians) as they make treatment decisions. While most cases of Chiari related syringomyelia are treated surgically, this suggests that some patients who fit a specific profile may be able to avoid surgery with careful monitoring.

Source: Natural history of Chiari I malformation-syringomyelia: longitudinal cohort study. Yuan C, Du Y, Yao Q, Zhang C, Zhang L, Liu Z, Wang K, Duan W, Wang Z, Wang X, Zeng G, Wu H, Chen Z, Heiss JD, Guan J, Jian FZ. J Neurol Neurosurg Psychiatry. 2025 Jul 17:jnnp-2025-336023. doi: 10.1136/jnnp-2025-336023. Online ahead of print. PMID: 40675800

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