

Topics: Stem Cell Therapy, Syringomyelia; Orofacial Symptoms; Chiari Prevalence

Conquer Chiari's monthly research updates highlight and summarize interesting publications from the medical literature while providing background and context. The summaries do contain some medical terminology and assume a general understanding of Chiari. Introductory information about Chiari, plus many more research articles, can be found at www.conquerchiari.org.

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 A research group from Spain has demonstrated that injecting a certain type of stem cells directly into a post-traumatic syrinx actually reduces the size of the syrinx and provides some improvement in sensation and function. Mesenchymal stromal cells (MSC) are a type of stem cell which is cultivated from the person who will in turn receive them as treatment and have previously been shown to provide some improvement to spinal cord injury patients. In this study, the researchers were investigating whether the presence of a syrinx – 1%-4% of spinal cord injury patients develop post-traumatic syringes – limited the effectiveness of the MSC treatment due to the large physical cavity that exists. Specifically, they injected 300 million stem cells mixed with plasma directly into the syringes of 6 adult men who had been living with their spinal injuries for 5-27 years. They then assessed the impact of the MSC injection across a number of measures, such as sensation, pain, bladder and bowel function, and MRI. They found the stem cells did provide some modest improvement across many of the measures, although statistical significance was difficult because only 6 people were tested. There were improvements in sensation, pain, bladder issues, and overall spinal function. But the real surprise came when they measured the syrinx size on MRI. They found a significant reduction in the syringes, especially the width. The scientists admit they cannot explain how this happens from the stem cell injection, and also point out that as the syringes shrank in size they often revealed a thinning of the spinal cord tissue which has previously been a predictor of a poor response to stem cell therapy. From this, they stress the importance of administering this type of therapy as early as possible. This type of stem cell therapy is no doubt expensive and heavily regulated, but it would be fascinating to see if it is also successful in shrinking Chiari related syringes and improving function.

SOURCE: *Cell therapy with autologous mesenchymal stromal cells in post-traumatic syringomyelia. Vaquero J, Zurita M, Rico MA, Aguayo C, Fernandez C, Rodriguez-Boto G, Marin E, Tapiador N, Sevilla M, Carballido J, Vazquez D, Garcia-Olmo D, Guadalajara H, Leon M, Valverde I; Neurological Cell Therapy Group From Puerta De Hierro-Majadahonda Hospital. Cytotherapy. 2018 May 18.*

 An often-overlooked aspect of Chiari symptomology received some attention recently when a team from Brazil looked at the orofacial symptoms of adult Chiari patients. In a small study, four men and two women were given a simple, but widely used questionnaire to assess orofacial issues such as jaw pain, trouble chewing, and clicking. Interestingly, common Chiari symptoms such as headache, neck pain, and ear pain can also be associated with orofacial and temporomandibular problems, so the assessment also asked about those. The researchers found that orofacial problems were indeed fairly common, including:

- Half of the patients reported pain or problems opening/closing the mouth
- Half reported pain or weakness when chewing
- 2 reported difficulty moving the jaw forward or sideways
- 5 reported headaches (4 had had decompression surgery), 4 neck pain, and 2 ear-pain

Despite the self-reports of pain and difficulty with opening the mouth, the researchers found that the patients were able to open their jaw to a normal distance. The authors also point out that the orofacial symptoms of Chiari can be mistaken for temporomandibular joint disorder (TMD) and highlight patients who were treated unsuccessfully with a dental approach because the problems were likely due to the underlying Chiari. It is not clear how many clinicians assess orofacial symptoms when evaluating Chiari, but it is clear that it is an area that deserves more focus and research.

SOURCE: *Orofacial clinical features in Arnold Chiari type I malformation: A case series. de Arruda JA, Figueiredo E, Monteiro JL, Barbosa LM, Rodrigues C, Vasconcelos B. J Clin Exp Dent. 2018 Apr 1;10(4)*

 Determining how many people have Chiari is difficult, partly because it depends on how it is defined. Using the classical, radiographic definition of 5mm or more of tonsillar herniation results in a huge number, with some studies showing that up to .9% of US adults fit this criterion. However, the overwhelming majority of those people will never experience any Chiari related symptoms, so this is not a particularly useful number. Previously, Conquer Chiari has used the number of decompression surgeries performed annually to estimate that as many as 1 in 1,000 people in the US have symptomatic Chiari, but this is based on a number of assumptions. There have been a couple true epidemiological studies published outside of the US, namely Japan and New Zealand, but it is not clear how applicable those are to the population as a whole. While Chiari appears to affect people of all races, there are indications that the prevalence of Chiari does vary from population to population and that it is not as prevalent in Asian countries. Recently, a careful study of the population of the Republic of Tatarstan in Russia (population 2.9 million) found the overall presence of Chiari in adults, defined as at least 2mm of herniation with Chiari type symptoms to be 30 in 100,000. Interestingly, the gender split was nearly 50-50 between men and women, which is much different than in the US. It is also very interesting to note that one-third of the group had tonsillar herniation of less than 5mm, the classic definition of Chiari, and that this group, similar to those with larger herniations, had smaller posterior fossa bones (and thus space) than controls. This means that if the medical community uses the 5mm definition as a hard and fast rule, they could exclude as many as one-third of symptomatic Chiari patients. One other item of note is that even within the geographic region that was studied, they found differences in prevalence between districts.

SOURCE: *Epidemiology of Symptomatic Chiari Malformation in Tatarstan: Regional and Ethnic Differences in Prevalence.* Bogdanov EI, Faizutdinova AT, Mendelevich EG, Sozinov AS, Heiss JD. *Neurosurgery.* 2018 May 21.

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