

Patients sometimes develop low pressure headaches as a result of spontaneous leaks of their cerebrospinal fluid (CSF), but it's not clear why those leaks happen. Three doctors studied 18 patients to find out if a connective tissue disorder (CTD) might be a cause. They noticed that some of these patients had indications of Marfan Syndrome, and others had signs of Ehlers-Danlos Syndrome. More specific tests that often can help make a CTD diagnosis did not help in this case, leading researchers to conclude that patients with a spontaneous CSF leak often can have a connective tissue disorder, but the signs may be very subtle. They also noted that problems with wound healing may occur as a result of a patient's CTD.

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## **Connective Tissue Disorders with Spontaneous Spinal Cerebrospinal Fluid Leaks and Intracranial Hypotension: A Prospective Study.**

### **CLINICAL STUDIES**

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#### **Abstract:**

**OBJECTIVE:** Intracranial hypotension attributable to a spontaneous spinal cerebrospinal fluid (CSF) leak is an increasingly recognized cause of postural headaches. The cause of these leaks is poorly understood, but it is likely multifactorial and may involve a primary connective tissue disorder. We undertook a study to estimate the contribution of systemic connective tissue disorders to the development of spontaneous spinal CSF leaks.

**METHODS:** We examined a group of 18 consecutive patients with spontaneous spinal CSF leaks for features of a connective tissue disorder.

**RESULTS:** The mean age of the 15 female patients and 3 male patients was 38 years (range, 22-55 yr). Seven patients (38%) demonstrated stigmata of a systemic connective tissue disorder, and three distinct types of disorders could be identified, as follows. 1) The association of spontaneous spinal CSF leaks and minor skeletal features of Marfan syndrome was noted for three patients. 2) Ehlers-Danlos syndrome Type II was noted for two patients. 3) Joint hypermobility associated with marked attenuation of the dorsal muscular fascia, precluding proper wound closure, was noted for two patients. In addition, isolated small-joint hypermobility was observed for five patients (28%). Slit-lamp ocular examinations, echocardiographic evaluations, histopathological examinations of skin biopsy specimens, and renal scanning did not reveal any other features of a systemic connective tissue disorder.

**CONCLUSION:** Findings suggesting connective tissue disorders are common among patients with spontaneous spinal CSF leaks, and manifestations may be subtle. A variety of disorders can be identified, probably reflecting genetic heterogeneity. Problems with wound healing may occur as a result of the systemic nature of the underlying connective tissue disorder.

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