

THE WAITING ROOM

THIS WAY IN

Clearing the Air on Cluster Headaches

They've been called more painful than broken bones, burns, or intense back pain from childbirth ("back labor"). They've been compared to having a red-hot poker stuck through your eye. They're so agonizing that they've even been called "suicide headaches."

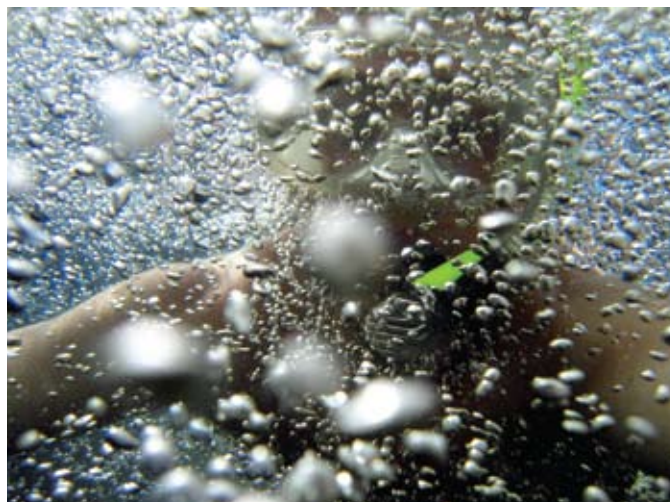
We're not talking about commonplace stress headaches, or the spinal headaches some people get as a result of spinal anesthesia, or even severe, chronic migraines. These "worst of the worst" headaches are called cluster headaches.

"Over the years, I've asked my patients what's worse: cluster headaches or passing a kidney stone, cluster headaches or delivering a baby, cluster headaches or any other kind of pain. They invariably say the cluster headache is the worst pain they've ever experienced," says Richard Lipton, M.D., professor and vice chair of neurology at Albert Einstein College of Medicine and director of the Headache Center at Montefiore Medical Center in New York, NY.

Cluster headaches typically affect one side of the head and do not switch sides. They usually occur in groups or "clusters" at the same time of day or night for several weeks, abate for a stretch of time, and then start up again. For example, a person may have two cluster periods each year separated by months of freedom from symptoms; during the cluster periods, he might experience one to three headaches a day for several weeks. An increased familial risk of cluster headaches suggests that they may have a genetic component. In addition, these headaches are more common in men than women and in smokers than nonsmokers.

Fortunately, cluster headaches are much less common than other kinds of chronic headaches, affecting about a million people in the U.S. every year. By comparison, for every one person with cluster headaches, about 100 might have migraines. But that relative rarity has a downside: Much less research has been conducted on cluster headaches than, say, migraines, and they've been harder to diagnose and treat.

"Most patients with cluster headaches are misdiagnosed," says Stephen Silberstein, M.D., professor of neurology and director of the Jefferson Headache Center at Jefferson University Hospitals in Philadelphia, PA. "Commonly, they're told instead that they have some type of migraine, or another pain syndrome



Oxygen therapy isn't the only treatment for cluster headache, but it's one of the fastest and most effective.

Cluster headache symptoms can include:

- ▶ Excruciating pain in or around one eye
- ▶ Tearing and redness in the affected eye
- ▶ Stuffy or runny nose on the affected side
- ▶ Drooping eyelid and decreasing pupil size on that side

called trigeminal neuralgia."

But now, a rigorous new study published in the *Journal of the American Medical Association (JAMA)* confirms the effectiveness of one of the key treatments

for cluster headaches: oxygen. The study may help both to raise awareness of the phenomenon of cluster headaches among general physicians and to ensure that this simple, effective treatment is covered by insurance.

"For years, specialists have believed that breathing high-flow oxygen is an effective treatment for cluster headache," says Dr. Lipton. "But the evidence from double-blind studies wasn't nearly as strong as it is now, with this new paper." The treatment involves breathing a high concentration of oxygen—in the study, 12 liters per minute—through a face mask for around 15 minutes at the onset of a cluster headache.

In the study, published in the December 9 edition of *JAMA*, investigators from the Headache Group at the University of California-San Francisco and the Institute of Neurology at the Na-

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tional Hospital for Neurology and Neurosurgery, Queen Square, London, compared oxygen therapy or placebo forced air in 109 adult patients during four cluster headache bouts.

After 15 minutes, 78 percent of the oxygen patients were pain-free, compared with 20 percent of the placebo forced-air patients. No significant adverse events were reported. “The authors did the world a great service with this really rigorous trial,” says Dr. Lipton.

You don’t need a huge supply of oxygen to treat cluster headaches. “Most patients have about one to three attacks per day, and a cluster of attacks lasts for about six weeks,” says Dr. Silberstein. “You usually only turn a tank of oxygen on for about 10 minutes at a time, so that’s about 36 hours’ worth of oxygen over a six-week period.”

Oxygen therapy isn’t the only acute treatment for cluster headache, but it’s one of the fastest and most effective. (Acute treatments are taken at the onset of an attack.) One of the most commonly used acute treatments for cluster attacks is injectable sumatriptan, a drug developed for migraine.

“Having options that will quickly abort the attack at the beginning of the cluster phase, without an injection, is very important. Oxygen is quite safe, relatively inexpensive, and works quickly,” Dr. Lipton says. And oxygen therapy has no known side effects, unlike sumatriptan, which has side effects that can include chest pain or tightness, neck pain, numbness—and more rarely, fainting, seizures, and allergic reactions.

“In addition to acute treatments, preventive treatments are also a therapeutic mainstay,” Dr. Lipton explains. “Preventive treatments are taken on a daily basis, whether or not a headache attack is present, to reduce the frequency of attacks. But preventive treatments can take a couple of weeks to kick in, and they are rarely 100 percent effective, making acute treatments extremely important,” he adds.

Until now, however, many insurance companies have declined to pay for oxygen therapy for cluster headaches, citing a lack of data. “Medicare won’t pay for cluster headaches, and neither will most insurers,” says Dr. Silberstein. “There’s no ‘indication.’ Because cluster headache is something of an orphan disease, just about nothing is ‘indicated,’ and in most of the insurance coding, it’s considered to be a migraine variant.”

The high profile of the *JAMA* paper may help to change that. “This is a publication that will be read by a wide range of physicians, and should both increase awareness of cluster headache among doctors generally, and help with reimbursement,” says Dr. Lipton. “When people with cluster headaches end up at my door, they often have been to four or five other doctors first. But now we may be able to increase the awareness of primary care providers and help see to it that insurance will pay for this very effective treatment.” —*Gina Shaw*

NEUROBICS

Back Words



Here’s a puzzle that exercises your ear for language. We recorded the names of a dozen famous movies, played them backwards, and transcribed the resulting nonsense words. For instance, *Star Wars* (s-t-ah-r-oo-oh-r-z) backwards is Zuroh Oorots, and *E. T.* (ee-t-ee) backwards is the same.

Can you identify the backwards movie titles below? Some are recent blockbusters, while others are older classics. Hint: Listen carefully to the actual sounds in each word. Sometimes the sounds don’t match the spelling. For instance, the long “i” in “lie” is really made of the two sounds “ah” followed by “ee,” so “lie” backwards is “yawl,” not “aisle.”

This puzzle was adapted from the book *The Playful Brain: The Surprising Science of How Puzzles Improve Your Mind*, by Richard Restak and Scott Kim, to be published this fall by Riverhead Books.

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|-----------------|------------------------|
| 1. ZAHJ | 13. TEA ALL ME ALL |
| 2. SHAM | 14. RAW TAVA |
| 3. ZROCK | 15. LA HEENA |
| 4. NARCH | 16. ARAB BACK |
| 5. E-CAR | 17. NO LUHMO |
| 6. JACE YAW | 18. SKIRT A METH |
| 7. OAK YOSS | 19. EAR OATSY OAT |
| 8. KINA TIGHT | 20. O MEAN NEED KNIFE |
| 9. SAFE ROCKS | 21. TEA ON CRAW DEATH |
| 10. POOH SCUD | 22. I’M A MOM |
| 11. TOE BOESH | 23. CROP KISS A ROUGE |
| 12. CHOKE JATES | 24. WANTS PILL A COP A |

ANSWERS ON P. 13

NEUROLOGY NEWS

Evaluation of Treatments for Muscle Cramps

A new American Academy of Neurology (AAN) guideline that evaluates treatments for muscle cramps advises against the use of the drug quinine, an anti-malarial drug that has both analgesic and anti-inflammatory properties. Although the guideline authors acknowledge quinine's effectiveness, they say its use should be avoided because of uncommon but adverse side effects. The guideline is published in the Feb. 23, 2010 issue of *Neurology*.

Only when the patient experiences disabling muscle cramps, has exhausted all other medication options, and has been made aware of the possible serious side effects can quinine treatment be considered, says lead guideline author Hans D. Katzberg, M.D., of Stanford University. Patients on quinine drugs should receive "frequent history and physical examinations, as well as routine blood work," Dr. Katzberg adds.

The guideline authors advise against the use of quinine because of the potential for toxicity, which can result in headache, dizziness, and hearing loss (cinchonism). Other more serious side effects may include a tendency to spontaneous bleeding (bleeding diathesis), and abnormalities—usually blood clots—that occur within the small blood vessels of the body.

Muscle cramps are temporary involuntary contractions of a muscle or a group of muscles that are sudden, forceful, and often painful and can last from a few seconds to minutes. A variety of neuropathic conditions, such as amyotrophic lateral sclerosis (ALS), peripheral neuropathies, and cramp-fasciculation syndrome (the uncontrollable twitching of muscles beneath the skin) are associated with muscle cramps.

The guideline authors evaluated treatments specifically for muscle cramps that have no known cause and those that oc-

cur as the result of neurological conditions, such as neuropathy or ALS. The authors reviewed 563 potential articles and identified 24 that satisfied their inclusion criteria of prospective trials (studies where participants are enrolled in clinical trials and investigators subsequently follow up with them) that evaluated the efficacy of a specific treatment for muscle cramps as a primary or secondary outcome. (The "primary outcome" of a study is the one the investigators are most interested in measuring; a secondary outcome is one that is of lesser interest to the investigators, but still of interest.)

Naftidrofuryl oxalate, diltiazem, and vitamin B complex can be considered possible treatments for muscle cramps, according to the AAN guideline. Naftidrofuryl oxalate is a drug that may enhance utilization of oxygen and glucose in peripheral vascular disease; however, it is not available in the U.S. Diltiazem, a calcium channel blocker, slows the electrical current in the heart, which slows heart rate and normalizes heart rhythm. Vitamin B complex is a compound that contains many groups of vitamins the body requires in small amounts to remain healthy. However, further research is needed on the safety and effectiveness of all of these therapies.

"Any future studies should also include an assessment of the impact of cramps on the quality of life and non-pharmacological interventions in the treatment of muscle cramps," Dr. Katzberg says.

Dr. Katzberg advises that anyone who experiences persistent or disabling muscle cramps be evaluated by a health care professional to determine whether an underlying neurological or other medical condition could be causing the cramps. —Kierstin Wesolowski

Quinine should be avoided because of uncommon but adverse side effects.

NEUROLOGY ON THE 'NET

Neuropathy Caregiver Blog

Angela Macropoulos recently launched a blog, "The Circle of Care: Considering the Neuropathy Caregiver" to share her caregiving experiences—which are, in Angela's words, "routine, but sometimes feel like the world without end."

Angela's mother, Josephine Macropoulos, has chronic inflammatory demyelinating polyneuropathy, an autoimmune-related neuropathy characterized by progressive

weakness and impaired sensory function in the legs and arms. For six years, Angela has been assisting Josephine at every level, from dressing to money management to shuttling her to doctor appointments—all while working as a corporate attorney. She has also written about neuropathy for *I.G. Living* and *The New York Times* wire service.

On her blog, Angela shares the mundane, the frustrating, and the sometimes humorous sides of balancing caregiving and working. While the blog is

a forum for sharing her personal observations and insights, she looks forward to developing a neuropathy caregiver's online community.

You can follow Angela's experiences at neuropathycaregivers.blogspot.com.

You can also follow her on Twitter @elderdaughter or angelamac and on her Facebook page, **ElderDaughter**.

For information on Neuropathy Week (May 17-21, 2010) go to Resource Central, page 36.





QUICK TIPS

Epilepsy, Menstruation, and Ovulation

I had my first seizure—a grand mal—when I was five years old. When my parents found me, my lips were blue, my eyes were rolled back, and my entire body was shaking. I was diagnosed with encephalitis. The virus had traveled to my brain, causing me to go into a four-day coma. The doctors thought that when I came out of the coma I would probably have some degree of brain damage.

My parents sat by my bedside and prayed. On the fourth day, my father looked up to find my eyes wide open. The first thing out of my mouth was “Can I have McDonald’s fries?” I sustained no brain damage, but the encephalitis left scar tissue in my brain, resulting in epilepsy.

My seizures came only occasionally as a child, until my hormones started to change.

I learned from my doctor—Orrin Devinsky, M.D. professor of neurology, neurosurgery, and psychiatry at New York University School of Medicine and director of the NYU/Mount Sinai Comprehensive Epilepsy Center—that the reason for increased seizures around the time of ovulation and the premenstrual phase is thought to be primarily an increase in the ratio of estrogen to progesterone. Hormones don’t cause seizures, but they can influence when seizures occur.

According to Dr. Devinsky, “When seizures occur exclusively or predominantly around the time of menses or ovulation (catamenial epilepsy), which they do for many women, it is especially important to maintain good sleep and avoid alcohol—certainly less than two beverages per day—during those times. For some women, a temporary increase in medication can be helpful if their menstrual cycle is regular. The use of hormonal therapy such as progesterone to prevent seizures remains more of a potential than a proven strategy, and we lack data to show that progesterone is definitely effective or safe as a long-term treatment for epilepsy around the time of menses or ovulation. For women with catamenial epilepsy, even removal of the ovaries and womb does not stop seizures, so effective antiepileptic drugs and a healthy lifestyle remain our best approach to help control seizures.”

Another thing I learned over the years is that our doctors can only do so much. If we don’t help ourselves, then we can’t expect to get better. Here are tips that I have developed for managing epilepsy.

1 Track Your Seizures: I use a seizure diary to track my seizures on an ongoing basis. For every seizure I experience, I write down the type of seizure and when it occurred. This information can help give your doctor a better idea of where the seizures are coming from. You can find a good seizure diary at epilepsy.com/seizurediary.

2 Review Your Seizure Diary: I review my seizure diary each month to find patterns and ask myself questions. Did my seizures mostly happen in the morning, in the afternoon, or at night? Was I doing anything different during those times? Were most of the seizures focal, partial, or grand mal? Did my body swing to a certain side?

3 Track Menstruation and Ovulation: I keep track of my menstruation and ovulation on a wall calendar near my desk. I usually menstruate every 28 days, so I highlight the 28th day and circle four days before and four days after that day. I do the same for ovulation. During this time I’m careful where I go and what I do, because my chances of having a seizure are higher.

4 Maintain a Healthy Diet: Since I have been watching what I eat and exercising regularly, I have lost weight and my seizures have decreased quite a bit. According to Dr. Devinsky, “evidence suggests that regular exercise and stress reduction can improve seizure control for some people.”

5 Take Your Medication: This is the most important piece of advice I can give. Taking your medication on time is very important, especially during menstruation and ovulation.

6 Lower Stress: For me, stress can bring on seizures, especially during menstruation and ovulation. Therefore, I try not to let minor problems, past mistakes, or worries about the future upset me too much. I try to enjoy the moment and focus on the present.

7 Set Aside “Me” Time: Take some time to relax: Take a bubble bath, meditate, or read a book. A little quality time can help you wind down and remember to do the things that keep you healthy. You’d be surprised how many things can affect epilepsy—negatively and positively—so take some time to recharge.

8 Keep a Positive Attitude: Yes, you have epilepsy. But has the disorder changed you for the better in any ways? Have you become stronger as a person? Would you be as caring to others with conditions and disabilities? I think my disorder has made me a better person.

Stacey Chillemi is the author of *Epilepsy: You’re Not Alone* (Lulu.com, 2006).

NEUROBICS ANSWERS CONTINUED FROM P. 11

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|-----------------------|-------------------------------|--------------------------------|---|
| 1. ZAHJ = JAWS | 7. OAK YOSS = PSYCHO | 13. TEA ALL ME ALL = LIMELIGHT | 19. EAR OATSY OAT = TOY STORY |
| 2. SHAM = MASH | 8. KINA TIGHT = TITANIC | 14. RAW TAVA = AVATAR | 20. O MEAN NEED KNIFE = FINDING NEMO |
| 3. ZROCK = CARS | 9. SAFE ROCKS = SCARFACE | 15. LA HEENA = ANNIE HALL | 21. TEA ON CRAW DEATH = THE DARK KNIGHT |
| 4. NARCH = TRON | 10. POOH SCUD = DUCK SOUP | 16. ARAB BACK = CABARET | 22. I’M A MOM = MAMMA MIA! |
| 5. E-CAR = ROCKY | 11. TOE BOESH = SHOWBOAT | 17. NO LUHMO = HOME ALONE | 23. CROP KISS A ROUGE = JURASSIC PARK |
| 6. JACE YAW = ICE AGE | 12. CHOKE JATES = STAGE COACH | 18. SKIRT A METH = THE MATRIX | 24. WANTS PILL A COP A = APOCALYPSE NOW |