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HEALTH&SCIENCE

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TUESDAY, MARCH 11, 2014

EZ

GENDER ROLES

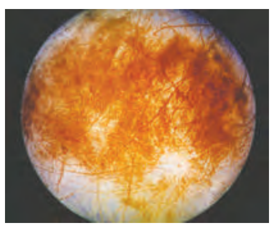
The Barbie effect
Playing with Barbies seemed to restrict little girls' ideas about what they could be; playing with Mrs. Potato Head did not. **E2**

QUICK STUDY

Anger and the risk of heart attack
Within two hours of an angry outburst, your risk of a serious cardiac problem is dramatically elevated. **E3**

SPACE TRAVEL

Is there life out there?
NASA would love to explore Europa, a Jupiter moon first spotted four centuries ago by Galileo. But can it be done? There are complications. **E2**



HEAVY DRINKING

Boomers, no binging
Older people who binge-drink tend to die earlier than those who consume the same number of drinks over a longer period. **E4**

Sleep too much?

ALAMY

Not a good sign.

Regularly spending more than 8 or 9 hours in bed may be bad for your health

BY CHRISTINA IANZITO

We get it: Sleep is good for us. The National Sleep Foundation regularly campaigns "to celebrate the health benefits of sleep," and experts have been boosting sleep's values as no less important than proper diet and exercise.

Insufficient sleep has been linked to stroke, obesity and heart disease. But sleeping too much may also be risky: It, too, is associated with a higher risk of heart disease and obesity, not to mention diabetes and depression.

So, how much is too much? And if you're sleep-deprived during the week, does sleeping 10 or 11 hours on Saturday and Sunday to catch up put you in any jeopardy?

Most experts say that a healthy amount of sleep for an adult is a regular seven to nine hours a night. And the operative term here is "regular," meaning the issue isn't the college kid who power-sleeps 15 hours on vacation to catch up from too much studying (or partying).

When scientists refer to "long sleepers," they're referring to people who consistently sleep nine or more hours a night, says Kristen Knutson, a biomedical anthropologist who focuses on sleep research at the University of Chicago's Department of Medicine.

"If you've been pulling all-nighters, by all means extend your sleep on the weekend if you can; try to catch up," Knutson says, "but if you're sleeping nine or 10 hours night after night after night for months on end . . . then we've got to understand why are you sleeping so much." You might be

SLEEP CONTINUED ON E4

Experts try to explain cases of paralysis

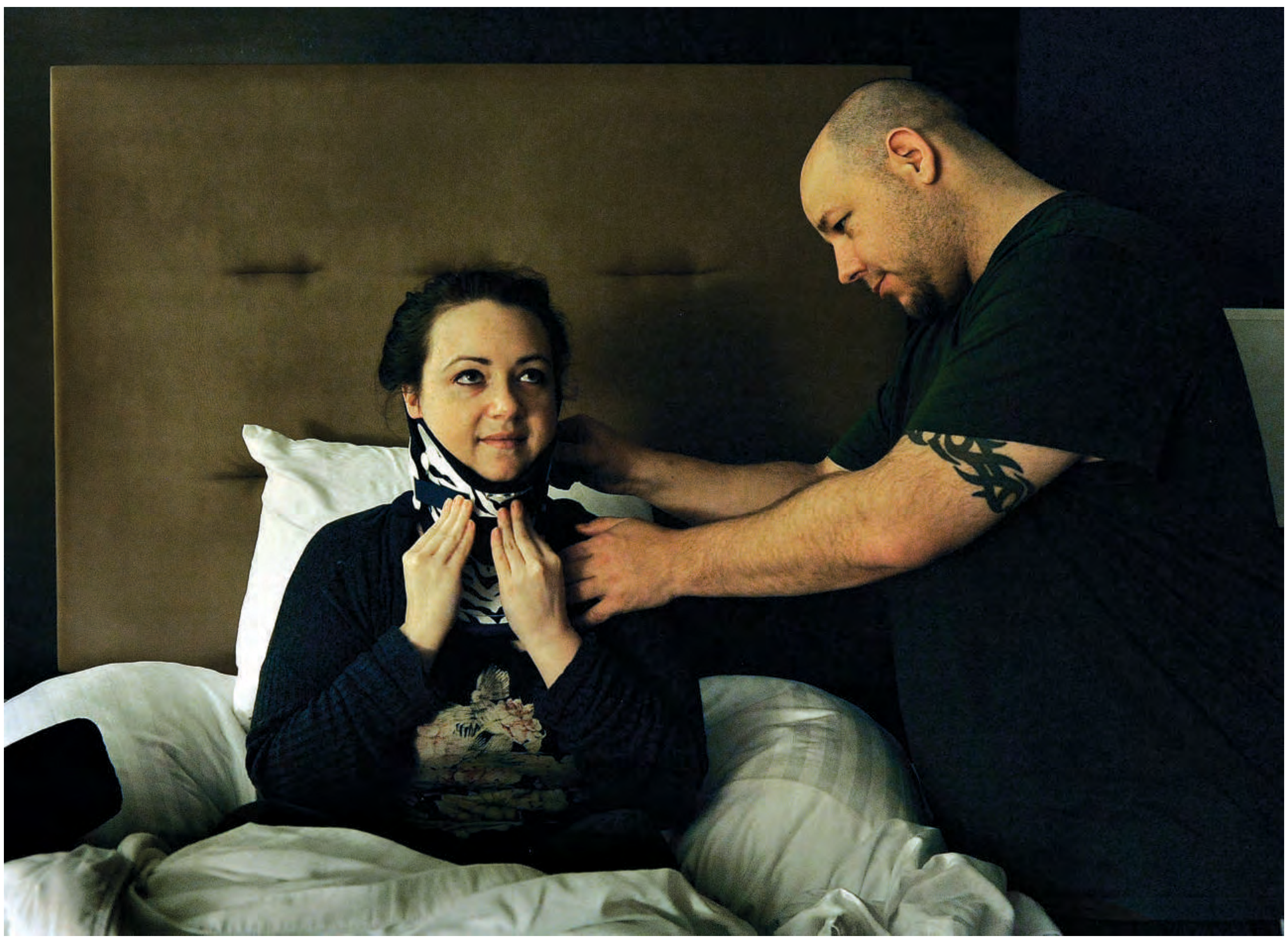
BY BRIAN PALMER

When a mysterious disease fells people — as happened in California recently, with as many as 20 children experiencing unexplained paralysis — teams of physicians and epidemiologists quickly mobilize. Perhaps you saw the movie "Contagion"? The idea is to find the culprit before it spreads but also to prevent public panic.

The investigation typically begins with a doctor reporting a sudden increase in patients with a particular disease or symptom to a state health department. It then falls to the government to determine whether the report is a false perception, a statistical quirk or a genuine surge.

The paralysis cases are a classic example: The symptoms are well known but their incidence appears to have spiked. Acute flaccid paralysis, the technical term for the symptoms observed in California, is something that many pediatricians have seen, and it has myriad causes. "I probably see one case like this every five years," says Keith Van Haren, the Stanford neurologist who is leading the hunt for an explanation for the paralysis reports. "Five cases in one year seems like an

VIRUS CONTINUED ON E6



PHOTOS BY MICHAEL S. WILLIAMSON/THE WASHINGTON POST

Kelly Koep adjusts her neck brace with the help of friend Josh Akins the day before surgery to counter the effects of a genetic disorder that kept her body from holding her bones together. Below: The day after the operation, surgeon Fraser Henderson checks to see if she is healing properly.

Stretched TO THE LIMIT

A genetic disorder left a woman's bones unable to support her body.

BY JEFF LEEN

Mildred Burke had one of the strongest necks in the world.

A champion pro wrestler in the 1930s and the first female athlete to earn $1 million, Burke was only 5-2, but her bulging neck measured 14 inches around. To entertain the crowd, she would lie on her back in the center of the ring and lift her body off the mat using only her head and neck, in what is known as a "bridge." She was featured in "Ripley's Believe It or Not" for performing 80 neck bridges at one time.

Mildred Burke's great-granddaughter has one of the weakest necks in the world.

Kelly Koep, 27, of Grant's Pass, Ore., has Ehlers-Danlos syndrome, or EDS, a genetic disorder that interferes with the synthesis of collagen. Once thought to be extremely rare, EDS was identified at the turn of the 20th century and comes in six forms that range from mild to severe.

Evidence suggests that the most common form, which often results in double-jointedness, occurs in one in 100 people; about one in 5,000 have more severe cases, which sounds like a nightmare out of science fiction: The ligaments in the body cannot hold the bones together.

Koep's is one of the most severe cases. Over the years, she has repeatedly dislocated elbows, knees, shoulders and — four times — her jaw. "I've always been that kid who could do weird, contortionist sorts of things," she said. "I can basically fold my back in half and rest my shoulder blades on my butt."

More recently, the increasing instability in her spinal column required her to wear a neck brace and to use a wheelchair. She got up only twice a day, to use the bathroom. There was concern that her loose vertebrae might be compressing the arteries in her neck, risking stroke.

"It felt like my head was so full of pressure that it was going to explode from

SPINE CONTINUED ON E5

Correcting the symptoms through surgery has given her new hope.

BRIAN SNYDER/REUTERS

Some people are repelled by the idea of cooking lobsters alive or the practice of tearing claws from live crabs before tossing them back into the sea.

If you were a crustacean, would you feel any pain?

BY TAMAR STELLING

A scientist and a seafood chef walk into a bar. "We have a mutual interest," says the scientist. "I study crustaceans and you cook them." But the chef wanted to know just one thing: Do the animals feel pain?

Robert Elwood had been working with crabs and prawns for the better part of three decades when Rick Stein confronted him with this question in a pub on the coast of Northern Ireland. Elwood was stumped. "It was the first time I ever considered the question," he says.

Although some people are horrified by the idea of cooking lobsters alive or the practice of tearing claws from live crabs before tossing them back into the sea, such views are based on a hunch. We know very little about whether these animals — or invertebrates in general — actually suffer. In Elwood's experience, researchers are either certain the animals feel pain or certain they don't. "Very few people say we need to know," he says.

The global food industry farms or catches billions of invertebrates every year. But unlike their vertebrate cousins,

PAIN CONTINUED ON E6

Are invertebrates able to feel pain?

PAIN FROM E1

they have virtually no legal protection. "Early on in my career I realized that when the law speaks of animals, it does not mean invertebrates," says Antoine Goetschel, an international animal law and ethics consultant based in Zurich. "As long as the common opinion is that invertebrates do not suffer, they are out of the game."

Pain vs. reflex

Pain is an awkward thing to test. It can't be measured directly or pointed at; it's not even easy to define. How can we tell when an animal is suffering? We have come a long way since Descartes, who argued that all non-human animals were merely automata, without self-awareness and incapable of feeling. But much of what we think we know still involves a lot of guesswork.

So how do we answer Stein's question? Elwood has been looking for ways to do so since running into Stein eight years ago. For a start, arguments by analogy are silly, he says. "Denying that crabs feel pain because they don't have the same biology is like denying they can see because they don't have a visual cortex."

Elwood and his colleagues at Queen's University Belfast are instead tackling the question by looking at how these animals behave. Most organisms can respond to a stimulus that signals a potentially harmful event. Special receptors called nociceptors — which sense excessive temperatures, noxious chemicals or mechanical injuries such as crushing or tearing — are found throughout the animal world, from humans to fruit flies. When a parasitic wasp jabs its egg-laying ovipositor into a fruit fly larva, for example, the larva senses the needle and curls up, which can make the wasp pull out.

But when an animal responds to something we would consider painful, it does not necessarily mean the animal is in pain. The response might be a simple reflex, where signals do not travel all the way to the brain, bypassing the parts of the nervous system connected with the conscious perception of pain. When we scald our hand, for example, we immediately — and involuntarily — pull it away. Pain is the conscious experience that follows, once the signals have reached the brain. The key for Elwood was to look for responses that went beyond reflex, the crustacean equivalents of limping or nursing a wound.

He started with prawns. After so many years of working with them, he thought he knew what to expect, which was that he would see nothing more than reflex reactions. But to his surprise, when he brushed acetic acid on their antennae, they began grooming the treated antennae with complex, prolonged movements of both front legs. What's more, the grooming diminished when local anesthetic was applied beforehand.

He then turned to crabs. If he applied a brief electric shock to one part of a hermit crab, it would rub at that spot for extended periods with its claws. Brown crabs rubbed and picked at their wound when a claw was removed, as it is in fisheries. At times the prawns and crabs would contort their limbs into awkward positions to reach the injury. "These are not just reflexes," Elwood says. "This is prolonged and complicated behavior, which clearly involves the central nervous system."

He investigated further by placing shore crabs in a brightly lit tank with two shelters. Shore crabs prefer to hide under rocks during the day, so in this situation they should pick a shelter and stay there. But giving some of the crabs a shock inside one of the shelters forced them to venture outside. After only two trials, the crabs that had received shocks were far more likely to switch their choice of shelter. "So there is rapid learning," Elwood says, "just what you would expect to see from an animal that experienced pain."

Finally, Elwood looked at how the need to escape pain competed with other desires. For humans, pain is a powerful motivational driver, and we go to great lengths to avoid it. But we also can override our instincts and choose to endure it if the rewards are great enough. We suffer the dentist's drill for the long-term benefit, for example. What would a crustacean want badly enough to make it endure pain?

For hermit crabs, it turns out to be a good home. These animals take up residence in abandoned seashells, but they can be made to give up their home if given a shock inside the shell. Elwood found that the likelihood of a hermit crab's dumping its shell when given a shock depends not only on the intensity of the shock but also on the desirability of the shell. Crabs in better shells took bigger shocks before they were willing to move out. This suggests that the crabs are able to weigh different needs when responding to the noxious stimulus. Once again, this behavior goes far beyond reflex, Elwood says.

JUNG YEON-JE/AGENCE FRANCE-PRESSE VIA GETTY IMAGES

A couple eats a live octopus during an event to promote a food festival in Seoul. An evolutionary neurobiologist in Texas has found that octopuses show much of the pain-related behavior seen in vertebrates.

The squid question

And it is not just crustaceans. Robyn Crook, an evolutionary neurobiologist at the University of Texas Health Science Center in Houston, is asking many of the same questions of cephalopods, such as squids and octopuses. "We are learning things we never expected to find," she says.

Crook and his colleagues have only recently shown that cephalopods have nociceptors at all. She also has found that octopuses show much of the pain-related behavior seen in vertebrates, such as grooming and protecting an injured body part. They are more likely to swim away and squirt ink when touched near a wound than elsewhere on their body.

Squids, though, may feel pain very differently. Shortly after a squid's fin is crushed, nociceptors become active not only in the region of the wound but across a large part of its body, extending as far as the opposite fin. This suggests that if it feels pain, rather than being able to pinpoint the location of a wound, an injured squid may hurt all over.

Crook is not certain why this would be. But it makes sense from a squid's point of view, she says. Unlike an octopus, a squid's tentacles can't reach many parts of its body, so it couldn't tend a wound even if it knew where the injury was. Squids also have a fast metabolism that forces them to stay on the move and keep hunting. An all-over heightened sensitivity may keep a squid generally more alert and wary. For example, Crook has found that an injured squid will be more sensitive than others to touch and visual stimuli. "Its long-term behavior changes," she says. "This fulfils one important criterion for pain."

If not the backbone . . .

Despite this work, the topic remains controversial. One concern is where to redraw the line if the backbone no longer marks a boundary. After all, roughly 98 percent of all animal species are invertebrates; Elwood and Crook may be only scratching the surface. The differences between octopus and squid show how diverse the experiences of the rest of the invertebrates might be, Crook says. A crustacean's neurons number in the low hundreds of thousands. If they feel pain, she says, what about fruit flies? They have a similar-size nervous system.

Fruit flies are known to have nociceptors, and it is likely that other insects do, too. Bees also respond differently to electric shocks given with and without anesthetic. And insects, generally, seem capable of learning to avoid noxious stimuli. But can they suffer?

Hans Smid, who studies the brains and learning behavior of parasitic wasps at Wageningen University in the Netherlands, dismisses the possibility. "I am absolutely convinced that insects do not feel pain," he says.

Like Elwood's, Smid's interest in pain began with a simple question. A few years ago, a visiting journalist was surprised at how casually Smid squashed a wasp that had escaped from its cage. Why hurt an animal you are so enthusiastic about, the journalist wanted to know.

Yet Smid is confident that insect behavior is best understood in terms of a relatively simple series of reflexes and innate responses. Unlike crustaceans, insects seem to have no pain-related behaviors. If an insect's leg is damaged, for example, it does not groom or try to protect the limb afterward. Even in extreme cases, insects show no evidence of pain. Imagine a praying mantis eating a locust, Smid says. With its abdomen opened up, the locust will still feed even while being eaten.

No gain, no pain

In terms of relative brain size, fruit flies and the parasitic wasps that Smid studies are the masterminds of the insect world. But neurons consume a lot of energy, and there is evolutionary pressure to keep brains as compact as possible. In short, there need to be good reasons to have enough brain for pain. Smid thinks that insects simply do not have the need. "I don't see the evolutionary advantage for insects to sustain such a complex system as emotion, of which pain would be just one component," he says.

Elwood agrees this is a useful way to frame the question. "From an evolutionary perspective, the only reason for pain that makes sense to me is that it enables long-term protection," he says. Pain may provide an animal with an additional, and memorable, means of focusing on a source of harm that helps it avoid it in future. If an animal's life span is not long enough to benefit from that — as is the case with most insects — then pain has no use. Similarly, some animals may simply be unable to avoid noxious stimuli in the first place. "Is a barnacle going to benefit from a bad experience?" Elwood says. "I doubt it."

Ultimately, we are up against the problem of consciousness. Like all subjective experience, pain remains private to each individual, leaving us only with educated guesses. But both Elwood and Crook have changed how they treat the invertebrates in their labs. They now use as few animals as possible and keep the potential for suffering to a minimum. And they are pushing others to do the same.

There are signs of change, too: Cephalopods at least now get some protection, in some parts of the world. "We are broadening our understanding of both pain and nociception," Crook says. "How can this not be interesting, even to the skeptics?"

This article was produced by New Scientist.

How researchers try to identify the cause of polio-like illnesses

VIRUS FROM E1

abnormality."

It's much harder to prove an abnormality than to perceive one, though. Surveys by the World Health Organization suggest that approximately one of every 100,000 children each year develops acute flaccid paralysis. It's not clear whether the rate is the same within the United States, but, since approximately 7.7 million kids younger than 15 live in California, 20 cases in a year might be within a normal range. A final determination on that question, however, will depend on whether the cases are related and whether they are clustered within a small geographic range. There is also a risk that clusters of cases can generate more such reports, especially once the media get involved.

"What you start looking for, you see," says Daniel Firkin, epidemiology branch chief in the division of viral diseases at the Centers for Disease Control and Prevention. Physicians usually don't report childhood paralysis, he explains, because the CDC doesn't require it. Now that they're on notice, however, more cases are going to be reported. It can be difficult to distinguish a surge in cases from a mere surge in reports.

Another challenge is determining which cases to include in the count. Establishing a case definition — a concise statement of what the patients have in common — prevents unrelated cases from leading investigators astray. According to Stanford's Van Haren, the working definition for the California cases is "paralysis with evidence of injury to the motor neurons in the anterior horn of the spinal cord."

That definition explains why the disease is being described as "polio-like." It's not simply that the children have become paralyzed. Several medical conditions can lead to paralysis. The autoimmune disease Guillain-Barrè syndrome and the inflammatory disease transverse myelitis, for example, can paralyze children. Poliovirus, however, has a specific profile, ending in paralysis, that sets it apart from most other illnesses.

MARTHA MENDOZA/ASSOCIATED PRESS

Jeff Jarvis with daughter Sofia, one of a handful of children with an unexplained illness that has left her arm paralyzed.

Polio is a classic fecal-oral disease. The virus usually spreads from one person's stool to another person's mouth, often through contaminated water or unclean hands. Since 95 percent of people who are infected with the pathogen never experience symptoms, polio victims rarely have reason to suspect they have come into contact with the disease; this is one reason that the eventual paralysis seems sudden.

In some patients, polio progresses to a brief bout of flulike symptoms, which has been observed in a few of the California patients. In a small minority of cases, the virus reaches the central nervous system. Poliovirus has proteins on its surface that match proteins on certain human nerve cells, as a key fits into a lock. Because the fit must be precise, poliovirus typically unlocks only the cells responsible for controlling muscles in the arms and legs — the motor neurons — in the front part of the spinal cord. Sensation usually remains intact in the paralyzed limbs, because polio can't enter the sensory nerves.

This clinical observation — paralysis of the limbs with little or no loss of sensation — probably rules out a large number of possible causes for the California cases, according to John Modlin, a physician working to eradicate polio at the Bill and Melinda Gates Foundation. Herpes simplex and varicella zoster, the pathogen responsible for chickenpox, are known to paralyze children in rare cases, but they usually affect sensation as well. The same is true for Guillain-Barré.

Since polio has been eradicated in the United States and the patients in California were vaccinated against it, the suspicion has fallen on other members of the enterovirus family, to which poliovirus belongs. Based on symptoms, doctors can probably rule out many of those viruses as well. Many enteroviruses have protein "keys" to other parts of the central nervous system. Some enteroviruses, such as the worrisome Enterovirus 71, which has caused outbreaks in Asia and other parts of the world, can inhibit breathing or heart rhythm by attacking respiratory or cardiac nerves. Those symptoms have not occurred in the California cases.

Despite all of these clinical clues, doctors can't know for certain what caused the paralyses until they catch the virus in the central nervous system. That's difficult, because enteroviruses typically remain in the spinal fluid for only a few days, or sometimes just hours. If the patient doesn't appear at the hospital immediately after the first signs of weakness, physicians have to sample other areas of the body to find the disease. Enteroviruses remain in nasal and oral secretions for up to 10 days and can be found in the stool for several weeks. Two of the victims in California had Enterovirus 68, another relative of poliovirus, in their nasal secretions.

Since polio has been eradicated in the United States and the patients in California were vaccinated against it, the suspicion has fallen on other members of the enterovirus family.

That doesn't prove anything, because enterovirus can be present without causing paralysis. The probability is significant, though. Modlin, a globally recognized expert on enteroviruses, sets the odds that Enterovirus 68 caused the paralyses in the children where it was found at "well in excess of 80 percent." Of course, it's entirely possible that one virus caused some of the paralyses while other pathogens were responsible for other cases.

There's one important question remaining to be answered: If an infectious agent is responsible for the paralyses in California, is this an outbreak that puts other children in danger? The question has even reached Washington, where Sen. Barbara Boxer (D-Calif.) is urging the CDC to open an investigation.

At this point, there appears to be little likelihood of an epidemic, according to doctors investigating the cases in California. The children had no apparent contact with each other, and physicians are still attempting to determine whether they have anything else in common, such as travel or contact with certain animals. If an enterovirus is the cause, the disease is unlikely to spread.

Aside from polio and Enterovirus 71, few of the pathogens in that family have shown any ability to cause epidemics. The California cluster may just be a tragic and infrequent consequence of a fairly rare infection.

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