

Mysterious fevers of unknown origin: Could surgery be a cure?

A child spikes a high fever, sometimes as high as 104 or 105 degrees, and sometimes causing seizures. She's rushed to the emergency room, the hospital runs test after test, specialists are brought in, but no explanation is found.

Many families – though no one knows how many – go through this cyclical nightmare. The fevers seem to come like clockwork, aren't accompanied by any obvious symptoms and don't respond to antibiotics or fever reducers like Motrin or Tylenol. Instead, they vanish on their own after four to five days, only to return four to six weeks later.

A report in last month's *Archives of Otolaryngology and Head and Neck Surgery* finds that tonsillectomy (with or without removal of the adenoids) is almost always curative. No one knows why – the tonsils and adenoids show no evidence of unusual infection or other abnormality when pathologists study them. But desperate families are opting for surgery as a last-ditch measure and finding it to be life-changing.

The paper, describing 27 children treated at Children's Hospital Boston from 2004 through 2006, offers the largest reported surgical experience to date with this syndrome, currently known as PFAFA (periodic fever, aphthous ulcers, pharyngitis and adenitis). Of the 27 children, 26 had complete fever resolution. Many had suffered cyclical fevers for years.

"Why taking out tonsils and adenoids works is unclear, but it works in almost every single kid," says Children's otolaryngologist Greg Licameli, MD, FACS, the paper's first author. "I tell parents, 'I don't know why this works, but it has a good chance of ridding your child of fevers.'"

Licameli has now seen 60 children with PFAFA, first described in the medical literature in 1987, and the findings continue to hold up. Given how many children he's seen in just five years, he thinks the condition isn't all that uncommon, though it is underrecognized and unknown to most pediatricians and otolaryngologists.

Licameli's introduction to PFAFA was a personal one: His first patient, described as a case report in the paper, was his own daughter, who at 19 months was getting fevers every three weeks. "There was nothing to break the fevers," he recalls. "She was seen by several experts at Children's. The workup was always negative."

Searching the medical literature for answers, Licameli found two small case studies in Europe, where doctors diagnosed PFAPA, removed the tonsils and the children got better, and decided to try this approach. It worked, with an immediate cessation of his daughter's fever cycles (she's now almost 6), and other doctors began referring patients to him.

The hallmark of PFAFA is high fevers of a cyclical nature – parents can often look at their calendars and predict what day the fevers will arrive. There are typically no other symptoms. Some children have sore throat, swollen glands and small mouth ulcers, but these symptoms aren't very specific and can be very subtle.

"On the face of it, it doesn't seem like a surgical problem," Licameli says. "The tonsils and adenoids appear normal, even when you study them pathologically, yet surgery is immediately curative."

Before undergoing surgery, patients are evaluated for any rheumatologic or infectious etiologies that can

also cause cyclical fever.

So what causes this condition, and why does surgery work? Licameli suspects the tonsils may harbor a chronic indolent infection to which the immune system is hypersensitive. He and his colleagues at Children's – including specialists in infectious disease, rheumatology and immunology -- plan to pursue these questions further.

In the meantime, they hope that increased physician awareness will allow children struggling with this condition to be offered surgery sooner.

Source: Children's Hospital Boston

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