

Autoinflammatory Diseases



DR. HAUS, RHEUMATOLOGY

KAWASAKI DISEASE AND THE END OF RHEUMATOLOGY AS WE KNOW IT

MAY 20, 2014 | JONATHAN HAUSMANN | [LEAVE A COMMENT](#)

This week, an international research team led by Xavier Rodó [published a fascinating study in PNAS](#) suggesting that Kawasaki disease is caused by an agent transported by wind from farms in Northeast China. This agent, possibly a fungal toxin, would be responsible for triggering an exuberant immune response in children, causing the typical manifestation of the disease: fevers, rash, conjunctivitis, “strawberry tongue,” enlarged lymph nodes, and swelling of the extremities. Untreated, Kawasaki disease can cause aneurysms of the coronary arteries, premature heart disease, and even death.

What I find so fascinating about this article is that it sheds light on the possible *cause* of a [rheumatic illness](#). As rheumatologists, one of the biggest challenges we face is not knowing the causes of most of the diseases we treat (that’s our dirty little secret!). Even though we use state-

of-the-art medicines, our understanding of disease is still in the Dark Ages.

Fortunately, we've had some progress. Rheumatic fever, for example, was found to be caused by *Streptococcus*, the same bug that causes Strep throat. We learned that treating Strep throat with antibiotics prevents rheumatic fever, likely the reason why rheumatic fever is now extremely rare in the United States.

In the 1970's, an epidemic of arthritis struck Connecticut, affecting many children. [Detailed research](#) showed that it was due to a bacteria, *Borrelia burgdorferi*, carried by a tick. To prevent disease, we advise people to use repellents and avoid tick-infested areas. If they develop Lyme, we offer effective treatments with antibiotics.

We have also made progress in understanding of some types of vasculitis (diseases that cause inflammation of blood vessels). Polyarteritis nodosa is often caused by hepatitis B virus, and cryoglobulinemic vasculitis is due to hepatitis C virus. [Cures](#) for these types of vasculitides can be achieved by eradicating the virus.

Is it a coincidence that several diseases that we considered to be "rheumatic" are now known to be caused by bacterial, viral (and perhaps) fungal elements? Not really, especially if we understand [evolutionary medicine](#). This often-overlooked field of study helps explain why humans, despite millions of years of evolution, are still vulnerable to disease. Two common reasons include pathogens (which are able to evolve faster than we can), and the mismatch between our bodies and our new environment (likely responsible for the obesity epidemic).

Unfortunately, most rheumatology research is conducted without an awareness of evolution. It seeks to find abnormalities of the immune system that cause disease, without first asking why any abnormality would exist in the first place. It tries to identify genes that make people susceptible for a disease, without asking how deleterious genes could be passed down through generations.

Fortunately, the winds of change may be near. [Interest in *P. gingivalis*](#) as a cause for rheumatoid arthritis continues to grow, and the role of the [microbiome in the development of rheumatic diseases](#) shows promise. With a better understanding of [why we get sick](#), we may uncover other environmental triggers responsible for the rest of the rheumatic diseases that we treat.

Gary Hoffman, a rheumatologist who studies vasculitis at the Cleveland Clinic, has said that understanding the *cause* of a disease is the "most crucial element." He [writes](#): "how empowering that knowledge is, especially when the etiological agent persists and perpetuates the process. In that setting, given adequate therapeutic interventions, we can even affect cures."

Scientific progress is [said](#) to occur through "paradigm shifts," or radical changes in our way of thinking, which abruptly transforms the field. Will a fungal toxin mark this change for rheumatology?