Regarding “Molecular diagnosis of nonaneurysmal infectious aortitis”

We read with great interest the case report by Kanemitsu et al showing the identification of bacterial species in infected, although not dilated, aortitis, which had been successfully treated surgically. Although the mortality rate of infected aortic aneurysm seems to have dropped in recent years, infected aortic aneurysm is still a diagnostic and therapeutic challenge. Considering nonelective surgery or non-operation increases the risk of aneurysm-related death, and early diagnosis of infected aortitis, followed by timely surgical intervention is, without doubt, beneficial to the patient.

In this case, however, Kanemitsu et al reported increased serum levels of immunoglobulin (Ig) G4 and high infiltration of IgG4-positive plasma cells in the adventitia of the descending aorta, both of which are characteristics of inflammatory aortic aneurysm and idiopathic retroperitoneal fibrosis. These disorders share similar clinical and histopathologic features and are both thought to result from an exaggerated inflammatory response to advanced atherosclerosis. In contrast to infected aortic aneurysm, inflammatory aortic aneurysm is presumed to be less susceptible to rupture, and corticosteroid therapy may be effective in controlling the perianeurysmal inflammation. In the absence of definitive diagnostic criteria for inflammatory aortic aneurysm, however, the selection of immunosuppressive therapy is sometimes a challenging option, and thus overindication should be avoided.

Results of blood culture can be negative in up to 50% of patients with an infected aortic aneurysm, and the prevalence of elevated serum IgG4 levels in patients with infected aortic aneurysm is not known. Despite these limitations, retroperitoneal fibrosis, rather than the infected aortitis, seems to be a more suitable diagnosis of the presented case, according to the serologic and immunohistochemical findings. Again, surgical intervention of the infected aorta, before it manifests apparent aneurysmal formation, may be an excellent choice for the management for this patient, especially considering that the aorta may show prominent enlargement over a short period of time and that arterial rupture can occur without aneurysm formation. Nevertheless, this case presentation raises fundamental questions:

1. Did this patient have both IgG4-related chronic periaortitis and aortic wall infection?
2. Do the authors think that IgG4-related periaortitis has a role in the development of bacterial infection-induced aortic wall remodeling, leading to aneurysmal formation, which is suggested in the pathogenesis of inflammatory aortic aneurysm?

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Reply

We thank Drs Ishizaka and Sakamoto for their interest in our article1 and are happy to reply to their comments. There is a close relationship between immunoglobulin (Ig) G4-related inflammation and fibrous sclerosing lesions. IgG4-related disease is clinically characterized by high serum IgG4 concentrations and is associated with multiple lesions in different organs. It can occur in the cardiovascular system and can manifest as an inflammatory abdominal aortic aneurysm (AAA). Scarring in the aortic wall sometimes extends into fibroadipose connective tissue around the aorta, and these cases are known as inflammatory AAAs associated with retroperitoneal fibrosis.

It is controversial whether a correlation exists between bacterial infected aortic disease and IgG4-related inflammatory aortitis. In our reported patient, histochemical analysis showed IgG4-related aortitis and, simultaneously, the molecular diagnostic technique of broad-range polymerase chain reaction and DNA sequencing revealed bacterial aortic wall infection. We do not have conclusive evidence that IgG4-related aortic inflammation has a positive role in the development of bacterial infection-induced aortic wall remodeling, leading to aneurysmal formation. We speculate that the bacterial wall infection was secondary to the IgG4-related inflammatory abdominal aortitis. Medial degeneration in the aorta is often due to medial laminar necrosis resulting from inflammatory processes. We speculate that bacterial infection extends to the aortic intimal wall and then infiltrates into the media, which results in a degenerative change due to IgG4-related inflammatory aortitis. The clinicopathologic characteristics of IgG4-related sclerosing disease in the aorta remain unclear.

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Endoleaks and the unending saga of a clever new terminology that has proved counterproductive

Incomplete aneurysm exclusion, both early and late, was identified as a unique complication of endovascular repair since its inception.1,2 The phenomenon was increasingly recognized in the mid-1990s as several authors noted the presence of “leaks” to signify persistence (or recurrence) of paragraft flow.3-5 White et al were first to describe the growing problem with clarity and coined a brand-new term: “We propose a preferable, novel terminology – endoleak – for this new phenomenon, which is associated only with endoluminal grafts.” The clever new term was received enthusiastically and adopted rapidly the world over.

For vascular surgeons, endoleak describes accurately the phenomenon of incomplete aneurysm exclusion. Unfortunately, everyone else in the medical community remains focused on the leak portion of the term, as it elicits deep-rooted mental images of a ruptured or rupturing aorta. While nuanced enough for vascular specialists, the differentiation intended by adding the prefix endo-to compose a wholly new word and concept failed to achieve its goal because it retained “leak” within the new term. The result: countless instances of unnecessary scare and anguish as physicians and patients become concerned – even panicked – by the thought of possible aortic rupture.

Nearly 15 years have passed and the endoleak saga continues to grow larger and increasingly problematic. Mainly, three factors combine to explain the current state of affairs: the rise of stent graft intervention as the new standard of care for most abdominal aortic aneurysm (AAA) patients; the prevalence of endoleaks after endovascular repair; and the enormous proliferation of computed tomography (CT) scan studies in the population at large.

Is there anything we can do now, can we extract the “leak” out of endoleaks? Is it too late to restore “saincy” with a new terminology that would serve us equally well to denote incomplete aneurysm exclusion but without any suggestion of a “leaking atheroma”? Several such terms have been used in published reports over the years, such as paragraft flow, persistent flow, incomplete exclusion, and the like. Among them, sac flow impresses me as the best candidate term to replace endoleak, as it is simple and precise, unique to aneurysms, and does not carry any hidden or overt implication of a potentially life-threatening situation. A contrast-enhanced CT scan (or ultrasound study) could be reported as showing evidence of sac flow or no evidence of sac flow. Moreover, sac flow could be further characterized as type I, type II, etc., in the exact same manner as endoleaks are classified today.

That said, I fully recognize mine is just a small voice in the wilderness. To resonate, a proposal of this kind will need to elicit enough interest from endovascular experts around the world who can, in turn, propel the discussion to higher levels. Furthermore, and ultimately, the major vascular and endovascular societies – and other stakeholders, regulators included – would have to become involved and embrace the cause. In the end, I feel strongly that resolution of this problem would result in significant benefit to our patients, and to us all.

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Reply
Thank you for the opportunity to reply to this letter. If Dr Criado is correct, and the sky is indeed falling,1 then it is highly unlikely that the solution lies in the simple substitution of a new neologism (“sac flow”2) for an older and well-tried neologism (“endoleak”).3

On the other hand, if the folk of Baltimore are truly facing an ongoing “unending saga,” including “unnecessary scare and anguish,” resulting in both “physicians and patients panicked,”4 then it may be best to initially look for a local solution, since such a state of panic does not appear to be widespread. I fail to perceive how the term “sac flow” could simply solve this problem, since by Dr Criado’s own logic, many outside the vascular community would now be forced to reinterpret new “deep-rooted mental images” of flowing blood and sacs. In addition, the term “endoleak” also defines the fact that an endovascular graft is present, whereas “sac flow” does not.

We proposed the term endoleak in 1996 precisely for the purpose of differentiating this new condition from the phenomenon of ruptured or leaking abdominal aortic aneurysm (AAA),3 and as an aid in providing a specific defined term and a classification system to help determine the long-term outcomes for the various types of endoleak that were being recognized in the early years of endovascular grafting.5,6 For example, this classification has helped enormously in showing that type II endoleaks do not require intervention in the high majority of cases.7 It is difficult to take at face value Dr Criado’s claim that “no other parts of the medical community have been able to comprehend the differentiation in the intervening 15 years, and that the simple use of a diagnostic term has elicited an “unending saga.”

A useful starting point for guiding the radiologists of Baltimore (or other regions of anguish) would be a series of recommendations for reporting post-endograft studies, which would always include a statement of the maximum size and integrity of an AAA or other aneurysm, as well as the presence or absence of endoleak and how it influences flow in the sac (“sac flow”). This system seems to work well for the rest of the world! It is difficult to see how a report that states that “type II endoleak is present, and is causing retrograde blood flow into the sac from a lumbar artery, without any evidence of AAA expansion or signs of AAA rupture” could be an inducer of panic. Such reporting guidelines are readily available from the American College of Radiology, and elsewhere.8 Also of concern in this letter is the implication that most US patients are