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More on Kartagener's syndrome and the contributions of Afzelius and A.K. Siewert

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The recent paper on Kartagener, Afzelius and immotile cilia syndrome [1] has evoked unexpected response. The first issue refers to Dr. Afzelius and Dr. Kartagener.

Dr. Afzelius offered some additional history. He actually had started his observations in 1959, in sea urchin sperm, where he described dynein arms and offered the hypothesis that the dynein arms make the microtubules slide relative to each other, causing the cilia and flagella to beat.

In 1974, he acted as opponent in the dissertation defense of a Danish colleague, Henning Pederson, who showed Dr. Afzelius his unpublished electron micrograph of an immotile human sperm tail lacking dynein arms. Afzelius asked him to publish this finding as soon as possible, which he did. Afzelius returned to Stockholm and contacted the andrologist, Dr. Rune Eliasson, and asked him whether he knew of any men who had stiff, immotile sperm tails; Eliasson knew of two brothers with this type of sperm, who gave ejaculates to be examined by electron microscopy.

Pederson and Afzelius published their papers *at the same time* in 1975, and Afzelius informed Eliasson that one of the brothers and a third unrelated man had immotile spermatozoa and *situs inversus* and problems with organs having ciliated epithelia. Pederson and Afzelius

succeeded in obtaining ciliated epithelia from them and showed the cilia lacked dynein arms; that was in 1976. Pederson died a short time thereafter. Dr. Afzelius is still active in 2004 at the age of 78.

Finally, Dr. Afzelius states that his first letter to Dr. Kartagener asking about male sterility was answered by Kartagener, who said that his patients were children. In a follow-up letter Dr. Afzelius told Dr. Kartagener he thought he had an explanation for the triad. Dr. Kartagener's daughter replied that he had died. Afzelius had intended to inform him about dynein arms; evidently this was never brought to Dr. Kartagener's attention.

The next concerns Dr. A.W. Siewert, who was referenced by Kartagener in 1933 as having published a case in 1904 of *situs inversus*, dextrocardia and bronchiectasis [2]. Though known to us, this case was not commented on in our paper since the focus of the paper was on the contributions of Kartagener and Afzelius and did not pretend to be an exhaustive review of "who named it first." Strangely enough, when looking up the triad syndrome, we found that there are over a dozen references to Siewert's paper on bronchiectasis, *situs inversus* and dextrocardia, but the syndrome is called Kartagener-Zivat or Zivert or Sivert; all of these papers were from Eastern Europe. Recently, Dr. Chris

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McManus in a letter to *The Lancet* [3] mentioned the mystery of Siewert: who was he and what happened to him?

It seems evident that the confusion on Siewert is due to not realizing that Siewert, although he published in German, wrote from Kiev, a city in imperial Russia with a large German population. Translation of the name Siewert from German to Russian in the Cyrillic alphabet and back to English seems to have produced confusion in the spelling of the name, such as Zivat or Zivert. Indeed, the 150th anni-

versary of the Faculty of the Kiev Medical Institute lists proudly Assistant Prof. A.K. Zivert as describing the congenital anomaly named the ZivertCartagener triad. (Note the spelling of Kartagener.)

In conclusion, the Siewert Kartagener syndrome came from German-speaking physicians who wrote in German but were themselves not German. Afzelius is correctly associated with them and indeed is Swedish.

Author's note Trying to write medical history papers is fascinating and frustrating—you are never finished.

References

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