Polysplenia or Left Isomerism?

Key words: polysplenia, asplenia, syndrome, isomerism, heterotaxy

(Intern Med 55: 555, 2016)  
(DOI: 10.2169/internalmedicine.55.5840)

To the Editor

Fukuda and colleagues described a patient with so-called “polysplenia syndrome” (1). Although frequently segregated on the basis of splenic anatomy, we would like to suggest that, from the cardiac point of view, splenic syndromes would be better assessed according to the morphology of the atrial appendages (2, 3). Most patients with multiple spleens, and all of those with left isomerism, have appendages with left morphology bilaterally. The counterpoint is that patients with right isomerism uniformly have appendages with right morphology bilaterally. There is, however, no specific arrangement of the abdominal organs that is significantly associated with either right or left isomerism. It is surely preferable nowadays to describe the abdominal organs based on their specific location, rather than creating uncertainty where none may exist by using alternative terms, such as “situs ambiguous”? Although patients with right isomerism usually show the absence of a spleen, they can have multiple spleens or even a solitary and normally located spleen. These findings go beyond mere anatomic curiosity, as they also portend important functional deficiencies. Functional asplenia, for example, may be present in those with solitary or multiple spleens (4, 5). Splenic anatomy should therefore not be used as a clinical indicator of the splenic function. The only justifiable assumption in this regard is that functional asplenia must exist when there is anatomic asplenia.

Fukuda and colleagues rightfully point to the association of isomerism with arrhythmias. As they emphasize, atrioventricular block is most frequent in those with left isomerism. A majority of these arrhythmias occur before 5 years of age, pointing to the importance of the findings in their patient. The authors comment that patients with left isomerism often die by 5 years of age, although they referenced this observation based on a significantly old study. Our previously reported analysis of over 5,000 patients with heterotaxy revealed that half of the subjects with left isomerism were still alive at 18 years of age. The use of optimal terminology in describing the overall constellation of symptoms thus conveys anatomic, functional and prognostic information. We propose that the evidence now supports a more effective description of splenic syndromes in terms of right or left isomerism, based ideally on knowledge of the morphology of the atrial appendages. If, however, the appendages are not visualized on imaging or at the time of cardiac surgery, the evidence at hand strongly supports the notion that the morphology of these structures can be inferred on the basis of associated congenital malformations (2, 3).

The authors state that they have no Conflict of Interest (COI).

Rohit S. Loomba 1 and Robert H. Anderson 2

References


1Division of Cardiology, Children’s Hospital of Wisconsin/Medical College of Wisconsin, USA and 2Institute of Genetic Medicine, Newcastle University, United Kingdom

Received for publication May 26, 2015; Accepted for publication June 8, 2015

Correspondence to Dr. Rohit S. Loomba, loomba.rohit@gmail.com

© 2016 The Japanese Society of Internal Medicine  Journal Website: http://www.naika.or.jp/imonline/index.html