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ORIGINAL ARTICLE

Thoracoabdominal Situs: A Practical Approach Accompanied by a Short History of Descriptive Terms

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Abstract Classification systems for thoracoabdominal situs have been the subject of numerous reports, yet confusion persists. This essay presents an abbreviated historical review of the currently used terms, briefly discusses some of the nomenclature controversies, and proposes a classification system that more closely adheres to the etymology of the descriptors, especially "heterotaxy."

Keywords Cardiac malformations · Heterotaxy · Medical history

Few topics engender more discussion, more passion between proponents of particular nomenclatures, and more potential confusion among clinicians than the description of variations in thoracoabdominal situs. Some situs descriptors have been in the medical literature for hundreds of years, and others were born in the twentieth century.

In 1987, Leonard Sacks and Ian Rifkin [6] wrote, "Problems in the understanding and classification of abnormal body situs have tormented clinicians and anatomists for years." Because many still share this sentiment, I offer a practical, clinical approach to classifying the variations in visceral situs that more closely follows the etymology of the descriptive terms.

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Short Historical Review

The first detailed reports of visceral arrangement abnormalities occurred more than 400 years ago. However, most credit the Scottish physician, Matthew Baillie, with the first comprehensive description of the anatomy consistent with situs inversus in 1788 [1].

Although Baillie's account was detailed, he did not use the Latin term "situs inversus" in his report. Nonetheless, the term did appear a few years later in a medical dictionary published in Germany in 1797 [5]. By the early nineteenth century, "situs inversus" appeared in German, French, Spanish, and English medical texts. The Greek-derived term "heterotaxia" (different arrangement) also began to be used in medical texts of multiple languages in the early nineteenth century. By the late nineteenth century, heterotaxia was used as a comprehensive term for visceral positions that differed from the usual and included situs inversus as well as other arrangements as subcategories [2].

In 1883, German physician Frederich Küchenmeister was likely the first to use another Latin term "situs solitus" to indicate the usual thoracoabdominal visceral arrangement [4]. From the seventeenth to the nineteenth century, authors of medical texts used other visceral situs descriptors, none of which is in general use today. For example, Küchenmeister's "situs solitus" persisted, but his lengthy Latin "situs viscerum totalis lateralis rarior, solito inversus" for situs inversus fortunately died out quickly.

Propelled by Maude Abbott and Helen Taussig, the interest in congenital heart disease increased substantially in the first part of the twentieth century, leading to more reports of cardiac defects associated with visceral situs abnormalities. In 1955, Biörn Ivemark [3] established a clear association between asplenia, thoracoabdominal situs

abnormalities, and cardiac malformations. Although Ivemark's name was applied to the constellation of abnormalities associated with asplenia, Ivemark's 1955 report also described patients with polysplenia.

After Ivemark's report, others developed the concepts of the asplenia syndrome, polysplenia syndrome, and isomerism of the atria and the lungs. In 1970, Lodewyk Van Mierop et al. [7] introduced another situs descriptor, "situs ambiguus" (uncertain situs), and assigned atrial isomerisms to this new category. Too many articles and textbook chapters on the subject of situs abnormalities have been written to be cited, yet even in 2010, published works using descriptive situs terms do so in confusing ways, albeit unintentionally.

Controversies and Nomenclature Confusion

From the last half of the twentieth century and through the first decade of the twenty-first century, the transglobal dialogue over the nomenclature of thoracoabdominal situs has continued. One of the issues has been whether atrial isomerism constitutes ambiguity or not, and the debate over this topic has been waged by the current great icons of anatomy, morphology, and pathology.

Another brisk colloquy has occurred over the use of the term "heterotaxy." In 2004, Stephanie Ware and John Belmont [8] wrote, "If right and left patterns of ordinarily asymmetrical structures are discordant, the resulting disorder is called situs ambiguus. A number of names have been used to describe this condition, including isomerism sequence, asplenia syndrome, Ivemark's syndrome, polysplenia syndrome, situs ambiguus, heterotaxia, partial situs inversus, and laterality sequence. A simplistic approach is to describe any arrangement of body symmetry that deviates from normal as heterotaxy, including both situs ambiguus and situs inversus."

Practical Points

Currently, two-dimensional echocardiography is the most common method used for anatomic diagnosis. Complementary tools include angiography, radiographic computed tomography, and magnetic resonance imaging. A clinical diagnosis is based on these methods, not on pathologic anatomy.

Segmental analysis is standard; and the first step in segmental analysis, either prenatally or postnatally, is to determine the visceral situs. The normal and abnormal arrangements of the liver and the major abdominal vascularity are easily determined by echocardiography. Although additional imaging methods will provide more anatomic details, echocardiography serves as the practical method of establishing visceral situs.

Particular attention is directed to the presence and position of the inferior vena cava relative to the abdominal aorta. An interrupted inferior vena cava accompanied by a large azygos vein posterior to the aorta strongly suggests features of bilateral left-sidedness or polysplenia syndrome. Bilateral right-sidedness, or asplenia syndrome, is suggested by an inferior vena cava that is anterior and on the same side as the abdominal aorta. If present, atrioventricular block is far more common in bilateral left-sidedness than in bilateral right-sidedness.

The morphology of the atrial appendages is rarely resolvable by echocardiography. Also, determination of thoracoabdominal situs is not directly dependent on the presence or absence of levocardia, dextrocardia, or mesocardia; types of atrioventricular connections; direction of ventricular looping; ventriculoarterial relationships; or additional intracardiac and extracardiac malformations. These malformations can occur with any situs arrangement, although their specific incidence does vary depending on the type of situs present.

Discussion

For more than 200 years, anatomists, morphologists, pathologists, and clinicians have developed many descriptive terms to characterize usual and unusual variations in thoracoabdominal situs. Currently, congenital heart disease specialists from different institutions or regions of the country or the world, influenced by training or the writings of iconic figures, use a variety of nomenclature systems. Some use the modifier "syndrome," even if by dysmorphology criteria no syndrome exists. A good example is "heterotaxy syndrome," which can be translated "different-arrangement syndrome." Because heterotaxy is a compilation term for a wide variety of different situs arrangements, it is not surprising that confusion exists. In contrast, "polysplenia syndrome" and "asplenia syndrome" do represent distinguishable sets of malformations.

From the perspective of a clinician and medical historian who has been confused by situs terminology for years, I propose using the classification that I outline in Fig. 1. In my opinion, the term "heterotaxy" should adhere to its Greek origins. That is, it should refer only to "different arrangements" from the usual. Thus, heterotaxy should be used as a comprehensive descriptor encompassing situs inversus, bilateral left-sidedness, bilateral right-sidedness, and situs ambiguus. Furthermore, it seems more logical to use "sidedness" rather than atrial isomerism because "sidedness" relates better to laterality than atrial isomerism.

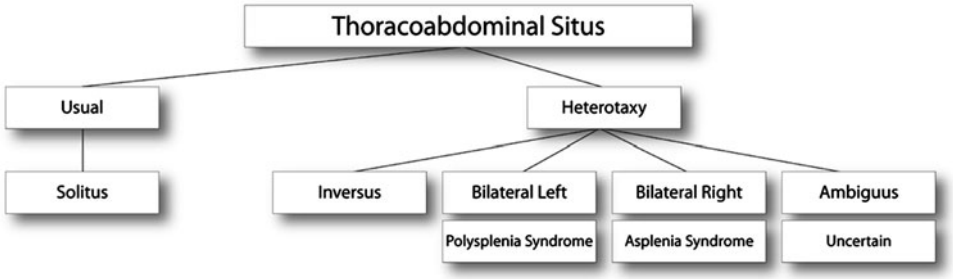

Fig. 1 Proposed classification of thoracoabdominal situs and its variations

Additionally, perfect atrial isomerism rarely exists; nor can the morphology of atrial appendages be determined easily by echocardiography. Within bilateral left- and right-sidedness, the terms polysplenia syndrome and asplenia syndrome respectively appear. Situs ambiguus should be reserved for arrangements that remain uncertain.

I do not believe that this proposal will end the confusion surrounding the description of abnormal thoracoabdominal situs. Nor do I believe it will be accepted by any number of anatomists, morphologists, pathologists, or clinicians. Nevertheless, I hope it will generate interest and debate.

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References

1. Baillie M (1788) An account of a remarkable transposition of the viscera. Trans R Soc Lond 78:350–363
2. Charpentier A (1887) Cyclopaedia of obstetrics and gynecology: a practical treatise on obstetrics, vol 3. Wood and Co, New York City, pp 283–284
3. Ivemark B (1955) Implications of agenesis of the spleen on the pathogenesis of cono-truncus anomalies in childhood: analysis of the heart malformations in splenic agenesis syndrome, with fourteen new cases. Acta Paediatr 44(Suppl 104):7–110
4. Küchenmeister F (1883) Die angeborne, vollständige seitliche Verlagerung der Eingeweide des Menschen: (situs viscerum totalis lateralis rarior, solito inversus); für Aerzte und Studirende. Barth, Leipzig
5. Ploucquet WG (1797) Initia bibliothecae medico-practicae et chirurgicae realis sive Repertorii medicinae practicae et chirurgiae, vol 8. U-Z. Cotta, Tubingae, p 414
6. Sacks LV, Rifkin IR (1987) Mirror image arrangement of the abdominal organs with a left-sided morphologically normal heart. Br Heart J 58:534–536
7. Van Mierop LH, Eisen S, Schiebler GL (1970) The radiographic appearance of the tracheobronchial tree as an indicator of visceral situs. Am J Cardiol 26:432–435
8. Ware SM, Belmont JW (2004) ZIC3, CFC1, ACVR2B, and EGAB and the visceral heterotaxies. In: Epstein CJ, Erickson RP, Wynshaw-Boris A (eds) Inborn errors of development: the molecular basis of clinical disorders of morphogenesis. Oxford University Press, Oxford, p 300