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Editorial

Evolving Understanding of Shone Complex Through the Lifespan: What's in an Eponym?

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See article by Aslam et al., pages 253–259 of this issue.

Eponyms are a mixed blessing. Most problematically, the name itself has no logical relationship to the diagnosis or constellation of characteristics. Use of an eponym, however, often indicates a particularly instructive, creative, or important association. Moreover, eponymous syndromes are more widely appreciated than their unbranded but otherwise equivalent cousins. It would be a safe bet that you know something about Churg-Strauss syndrome. We would need long odds to put our money on your familiarity with eosinophilic granulomatosis with polyangiitis. Who knew they were the same thing?

Some eponyms live up to their full potential. The word Eisenmenger might, to a naive audience, not bring to mind any aspect of the syndrome to which it refers. However, Eisenmenger syndrome exemplifies a useful eponym. Although it might be impossible to show causality, why else would there be essentially ubiquitous appreciation by non-specialists of such esoteric and complicated pathophysiology? But learning about Eisenmenger syndrome is far from a waste of time. Developing this specific conceptual understanding provides fundamental physiologic insights that apply to many diseases.

Other eponymous diseases might not carry such rich pathophysiologic treasure, but at least allow 'chunking' of universally linked features. Tetralogy of Fallot is much easier to remember and communicate than a list of the component lesions or description of the underlying developmental cause. It is true that the 4 components are not independent or equivalent (eg, right ventricular hypertrophy is due to subpulmonary stenosis), and that the term does not even refer to the primary lesion, anterior malalignment of the infundibular septum. At least it is clear what one means by tetralogy of Fallot, and the existence of an eponym suggests an important single diagnosis and saves time.

Shone complex (or syndrome or anomaly) is a less sanguine eponym. Sometimes the term indicates the precise set of all diagnoses first described by Shone and colleagues in 2 patients (Fig. 1).[1] It might also be used to refer to a *forme fruste* of the complete constellation, some specific subset of 2 or more left-heart obstructive lesions, as it was in 6 of the 8 patients presented in the original report.[1] The association of multiple left heart obstructive lesions highlights a likelihood of overlapping or dependent developmental origins. It does not, however, illustrate a clinically actionable concept. And contemporary clinical decision-making is on the basis of the individual lesions and their combined effect, without specific consideration of whether or not Shone complex is present. Having multiple defects is worse than having one. From the patient's perspective, does having Shone complex mean anything more than having multiple left heart obstructive lesions?

Dr Aslam and colleagues in this issue of the *Canadian Journal of Cardiology* present a retrospective study of adults with Shone complex cared for at the Montreal Heart Institute.[2] Their analysis of patients cared for between 1982 and 2014 helps provide perspective on how we should think about Shone complex in adults. Complete Shone complex, recapitulating the full set of specific lesions in the original report (parachute mitral valve, supramitral ring, left ventricular outflow obstruction, and coarctation of the aorta), was exceedingly rare. There was only 1 case, or 0.02% of the 4189 patients cared for in this clinic. If anything, that should overestimate the adult population prevalence because more severe cases are more likely to be referred to this specialized centre. Incomplete Shone complex, as defined by the authors to consist of at least 1 left ventricular inflow tract lesion and at least 1 left ventricular outflow tract lesion, was more common, occurring in approximately 0.67% of patients. Of course, these figures might not parallel birth prevalence, due to attrition before adulthood and the consequences of referral patterns. It is also notable that there are differing definitions of what constitutes Shone complex in the literature, adding imprecision to any prevalence estimate. The current report used a mainly anatomic definition (eg, would include a patient with a supramitral

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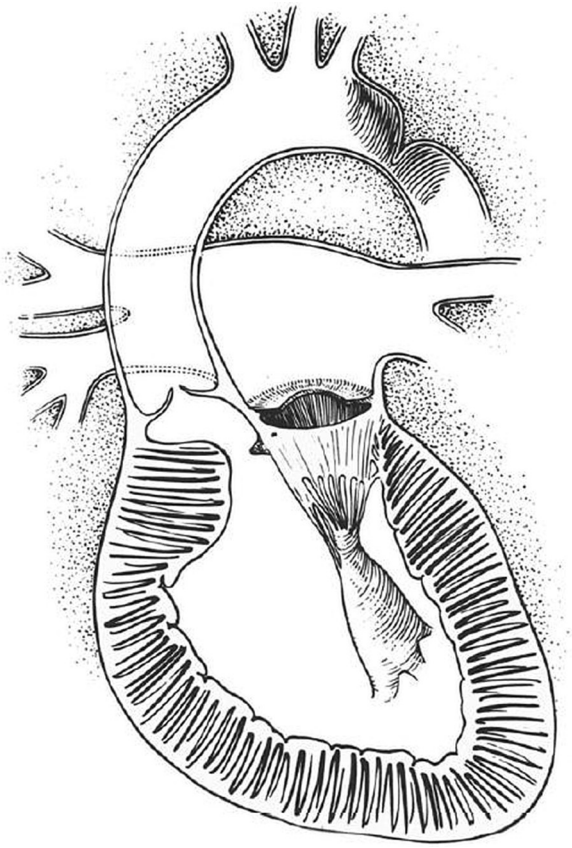

Figure 1. Diagram from the initial report of Shone complex illustrating the 4 obstructive anomalies: supravalvular ring of left atrium, parachute mitral valve, subaortic stenosis, and coarctation of aorta. Reproduced from Shone et al.[1] with permission from Elsevier.

ring and a bicuspid valve) whereas another excellent recent report on 121 children with Shone complex focused on those with actual left ventricular outflow obstruction.[3] There is no "correct" universally accepted definition of Shone complex. Despite their subtlety, such differences in definition complicate comparisons between studies.

Interestingly, only 11 of the 28 patients identified carried a diagnosis of "Shone complex" in the medical record despite receiving care at a referral adult congenital heart centre. This might partly be because of a more stringent application of the term by some cardiologists, as the authors suggest. It also could be related to the high prevalence of previous procedures and presence of prosthetic valves. A detailed review of distant historical documentation is often required to make a diagnosis of Shone complex in a newly referred adult patient who has had a prosthetic mitral valve replacement and coarctation repair. The clinical relevance of that diagnosis is less apparent than the direct implications of the intervening procedures; although it is best to know the type of congenital mitral lesion compelling previous mitral replacement, for providing good care now it is much more important to know the type of valve and how it is functioning.

The high (21%) prevalence of left superior vena cava (SVC) is also notable.[2] This presumably reflects a developmental association. Left SVC is not rare in general but is more common in patients with any type of congenital heart disease, and has been reported in the fetal echocardiography literature to be even more frequent among those with left-sided obstructive lesions.[4-6] Indeed, left SVC was present in 1 of the 8 cases from Shone's initial report.[1] Because of that context it is far less likely, although logically possible, that this observation reflects a survivor bias in which patients with a left SVC might have better survival to adulthood. The authors highlight the importance of this observation for surgical planning and for transvenous pacemaker or defibrillator lead placement. Of tangential interest, coronary sinus dilation related to left SVC can also rarely progress to cause left ventricular inflow obstruction.[7] In a patient with coarctation and bicuspid aortic valve, this acquired obstruction should not be confused with Shone complex.

The authors' data make a case, within the limitations of the study design, that the diagnosis of Shone complex might have real value above and beyond appreciation of the component lesions. These patients have a substantial burden of left heart failure and electrophysiological complications. We do not know empirically if morbidity is greater for patients with Shone complex than would be expected with a similar array of left-sided lesions (or with prosthetic valves) not meeting the criteria necessary to be labelled as Shone complex. There is, however, definitely substantial risk among those who do clear that bar, and the risk might be much higher than we usually attribute to coarctation or other isolated left-sided obstructive lesions. Additionally, a recent report on the clinical course of children diagnosed with Shone complex suggests pediatric outcomes are better than previously reported, foreshadowing a growing population of adults with this diagnosis.[3] Taken together, these data argue that we should be more proactive in recognizing the presence of Shone complex and more aware of the high risk of adverse outcomes in these patients as they age.

Disclosures

The authors have no conflicts of interest to disclose.

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