*Cardiology in the Young*

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# Richard Van Praagh—an appreciation

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## Obituary

**Cite this article:** Anderson RH and Houyel L (2025) Richard Van Praagh—an appreciation. *Cardiology in the Young* **35**: 2629–2632.
doi: 10.1017/S1047951125110214

Received: 26 September 2025
Revised: 8 October 2025
Accepted: 10 November 2025
First published online: 5 December 2025

**Keywords:**
Richard Van Praagh; segmental analysis; paediatric cardiology

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**Abstract**

With the passing of Richard Van Praagh in September of 2025, paediatric cardiology has lost one of its giants. With one of his greatest contributions, namely the segmental approach, he revolutionised the practice of paediatric cardiology. He and Stella, his wife and closest collaborator, developed the Cardiac Registry at Boston Children's Hospital and published more than 300 articles. Together, they trained many fellows, with endless generosity and dedication, and gave conferences all over the world, spreading their ideas and concepts. Richard will be sorely missed, but he has transmitted to us the desire and the tools to continue to build on his firm foundations.

With the passing of Richard Van Praagh in September of 2025, paediatric cardiology has lost one of its giants. During his long and fruitful life, there can be no question but that Richard has revolutionised the approach to the diagnosis of children born with congenitally malformed hearts. More than that, he has offered a means of how best to describe them. Prior to his innovations, it was the case that the more complicated lesions would be consigned to a category labelled "miscellaneous." Oftentimes, they would have no name. Subsequent to his initial publications, it was realised that, providing analysis started with a detailed analysis of the make-up of the different components of the heart, followed by a catalogue of all the lesions contained within the heart, nothing was beyond the realms of description and understanding. This was the essence of perhaps his greatest contribution, namely the segmental approach.[1] He did not make his achievements single-handedly. Throughout his long career, he collaborated widely. This was done not only with his direct clinical colleagues but also with the multiple fellows who studied and learned at his side. In his own words, when giving an interview to support the publication of his final book,[2] over which he had been toiling through his retirement, "My colleagues and I have published more than 300 scientific papers and 3 books. In all of this work, I have been assisted by paediatric cardiology fellows, cardiac surgery residents, and many wonderful friends and colleagues from all over the world." The initial interactions with his fellows were usually inspirational. They tended to colour their subsequent careers, as was certainly the case for the first author of this appreciation. His influence was equally important for those who did not necessarily see eye-to-eye, as in the case of the current second author. It is of note that one of his major collaborators, Aldo Castaneda, writing in a letter to a journal of cardiothoracic surgery, emphasised that his name "will eternally occupy a central place among the pioneers of congenital cardiac pathology."[3] Aldo also commented that "Richard was passionate about his work and consequently engaged not infrequently in intense arguments with his colleagues in paediatric cardiac pathology. Often these differences spilled over into, what I liked to call, transatlantic taxonomic warfare, mostly with Robert Anderson, a similarly opinionated British pediatric cardiac pathologist. Fortunately, these sometimes heated exchanges never succeeded in affecting their mutual professional respect or their friendship."[3] It is a privilege for us now jointly to endorse the validity of this insightful comment.

Richard was born in 1930. He arrived in April in London, Ontario, just a few months after the birth of Aldo. His initial training was in Toronto, where he came under the influence of John Keith. Keith had edited one of the first significant textbooks of paediatric cardiology, along with Richard Rowe, at the time working at Johns Hopkins in Baltimore, where Richard also undertook part of his training, and Peter Vlad, who was the chief of paediatric cardiology in Buffalo, in upstate New York. It was in Buffalo, in 1962, where Richard made perhaps his most important observation. It was there that he met and married Stella Zacharioudaki (Figure 1). Stella had been born in Crete and, like Richard, was training to become a paediatric cardiologist. Together they became a formidable team. In 1965, by which time Richard had conducted the research that formed the basis of the segmental approach,[4] he and Stella were invited to join the staff of Children's Hospital in Boston. There, until their retirement in 2002, they developed the Cardiac Registry as a centre for advancing the understanding of the pathology of congenital cardiac disease (Figure 1). Many of his most important contributions, as he explained in a tribute to Stella after her untimely death in 2006, depended on her promptings and suggestions.[5] As one of us can attest, having been trained in the Cardiac Registry in 1991, the support provided by

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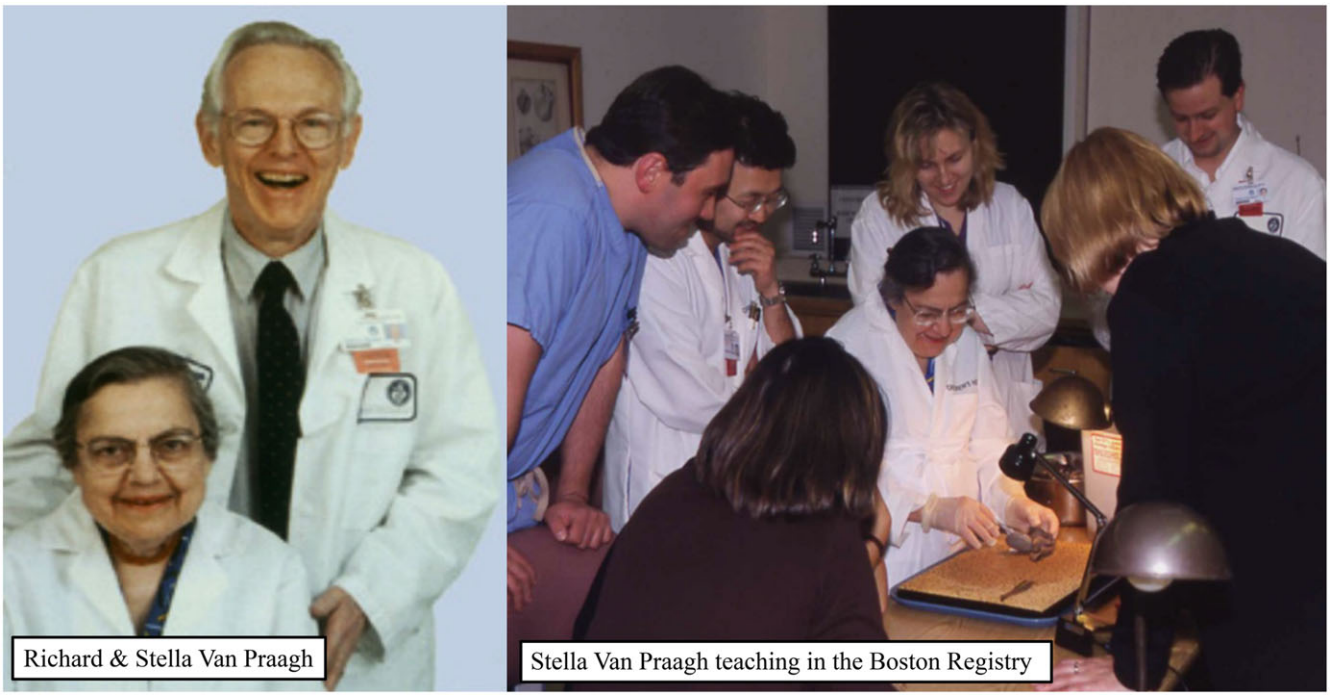



**Figure 1.** Left-hand panel: Richard and Stella Van Praagh. Right-hand panel: Stella teaching a group of fellows in the cardiac registry at Boston Children's Hospital.

Stella was also integral in providing the environment enjoyed by all of those who worked with Richard and Stella in the Cardiac Registry. Her baclava was both legendary and monumental, provided to the attending fellows with maternal solicitude. The major part of the teaching, provided with passion and endless patience, revolved around the specimens. The discussions taking place during the autopsies often extended over many hours, sometimes reaching into the first hours of the night. They served as the basis for worldwide dissemination of their concepts. Indeed, the enthusiasm and the generosity with which they shared their knowledge, as Lucile can testify, made her immediately feel the necessity, once returned to Paris, to transmit and increase this knowledge, but this time based on the meticulous observation of the heart specimens present in the Parisian collection. This was done in close collaboration with Richard, who always responded in long manuscript letters to her interrogations and thoughts, and by Stella, who did not hesitate over the subsequent years to spend hours with her over some chosen heart specimens, thus reinforcing her initial training. Richard and Stella also heartily opened their home to their fellows. This gave Lucile the immense privilege of essentially becoming part of their family, enjoying many precious moments of pure friendship over a good meal with excellent wines.

Their worldwide influence had already spread far and wide by the time that Robert, in 1974, was just beginning his own career. Richard had become a frequent visitor to London. Already, he had spent time with colleagues at Great Ormond Street looking at images that had been called in questioning the "loop rule" that had developed from his initial studies underscoring the segmental approach. Michael Tynan and Ian Carr had assembled the angiograms from over 100 patients with regular transposition. Richard's initial investigations have suggested that the location of the aorta, when abnormal, was a reliable indicator of the type of the associated ventricular loop. On studying the angiograms, with Richard making the decisions, the observers were surprised to find that, in about one-fifth of the datasets, despite the presence of a rightward loop, the aortic root was left-sided relative to the pulmonary trunk. This important information, however, although presented as an abstract at the annual meeting of the American Heart Association in 1969,[6] was never written up as a peer-reviewed publication. The subsequent realisation that such hearts with disharmony between the ventricular loop and the discordantly connected aorta represented a specific subset of transposition became an integral part of the ongoing collaborations between Lucile and Richard after Lucile had become a consultant at Marie Lannelongue Hospital in Paris (Figure 2).[7] But that was very much later.

Richard's curiosity was tireless. He was deeply convinced, since his first morphologic observations, that deciphering the mechanisms of cardiac development was mandatory for understanding the morphology of congenital heart defects. He was a pioneer in contributing to several experimental studies on human and animal embryos. Cardiac embryology was an integral part of teaching in the Cardiac Registry, leading to many passionate and thought-provoking discussions. Both Stella and Richard were remarkably open-minded, urging their fellows to question their teaching and to read authors with divergent opinions, permitting the fellows to make up their own minds. Both travelled all over the world, giving countless lectures, and always answering with great kindness every question of the audience, especially those of residents and young doctors.

Returning to the development of the segmental approach and the disagreements that subsequently arose, Richard was visiting London again in 1974 at the time that Robert, having just returned from a year working in Amsterdam, was setting up his own archive of congenitally malformed hearts at the Brompton Hospital. Richard was staying with Elliot Shinebourne, who had been responsible for recruiting Anderson to the Brompton. Whilst in Amsterdam, together with Anton Becker, Bob had studied a series of hearts with tetralogy of Fallot.[8] Their findings had raised questions as to whether the subpulmonary infundibulum was "too short, too shallow, and too narrow," as had been suggested by

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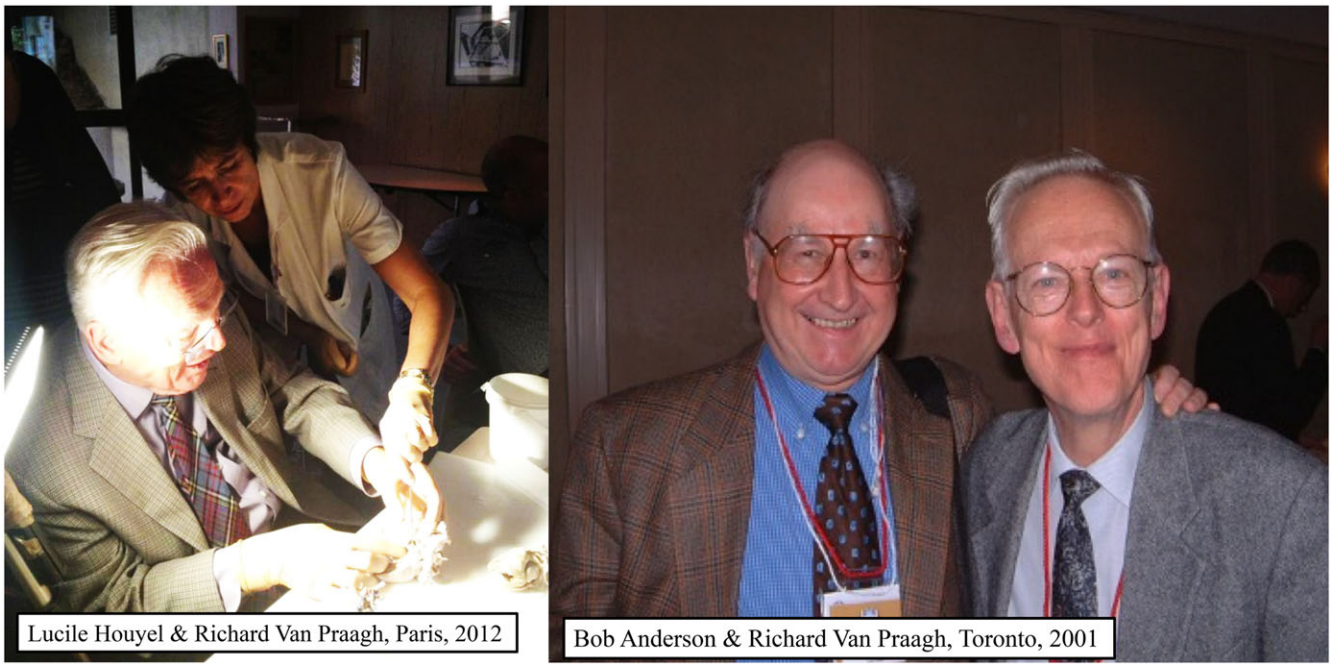



**Figure 2.** Left-hand panel: Richard demonstrating the particular features of an interesting specimen to Lucile in her own archive in Paris in 2012. Note that Richard is wearing a tie made of material showing the Anderson tartan! right-hand panel: Richard photographed with Robert Anderson during the Nomenclature Summit Meeting held in 2001 during the World Congress staged in Toronto.

Richard.[9] The potential differences resulted in animated and good-natured discussions between Bob and Richard, resulting in the eventual invitation for both Anderson and Becker to visit Richard and spend time working in the cardiac registry in Boston. The visit took place in 1976, during which time lengthy discussions seemed to have resulted in consensus with regard to the optimal way of assessing the congenitally malformed heart. In this regard, another of the insightful comments of Castaneda is pertinent. As Aldo commented, "I must confess that, not infrequently, I failed at first to recognize the essence of his arguments and only after further reflection managed to grasp the importance or validity of his ideas."[3] After their own further reflections, it remained the fact that, on their return to Europe, Anderson and Becker had still failed to grasp one of the basic tenets of Richard's philosophy. When producing the sequential modification of the segmental approach, the Europeans, in placing emphasis on the junctions between the segments, and subsequent to their discussions in Boston, had believed that they were doing no more than tweak the American concept. Even having received a lengthy 12-page criticism of their original draft, they still failed to appreciate that their new definition of "concordance" was sufficiently different from the stance formulated by Richard to raise significant objections from his side. It was not until decades had elapsed that the penny eventually dropped. Richard had based his concept on the notion of harmony and disharmony between the segments themselves, rather than on the way that the components of the segments were joined together. For Richard, irrespective of the fashion that the cavities of the components were in communication, or in some instances not in communication, the segmental combinations of {S,D,*} and {I,L,*} represented atrioventricular concordance, whereas the combinations of {S,L,*} and {I,D,*} were discordant. It was unfortunate, therefore, that the Europeans had chosen to designate hearts with double inlet ventricles, or the absence of one atrioventricular connection, as specific junctional variants, using the terms concordance and discordance to account only for the arrangements in which the atrial chambers were joined to their underlying ventricles in a morphologically appropriate or inappropriate fashion. It was the failure to grasp this simple fact that resulted in the subsequent schism in nomenclature emphasised by Castaneda as "transatlantic taxonomic warfare."[3] Perhaps the strife could have been avoided had the particular Europeans been aware that Richard was concerned with segmental, rather than cavitary, harmony as opposed to disharmony. But the damage had been done. Discussions, nonetheless, continued on the anatomy of many other lesions and many different segmental combinations. In all of these, the criticisms provided by Richard proved crucial in fine-tuning the multiple concepts being developed in London and Amsterdam. Of these, perhaps the most valuable was the justified criticism provided by Richard when the British consortium sought to disqualify the incomplete left ventricle from its morphological identity as a true "ventricle" on the basis that it lacked its inlet component.[10] As Richard and his colleagues pointed out, any component of the heart is best identified on the basis of its most constant component, which for the ventricular chambers was the apical trabecular part.[11] This notion is the basis of the "morphological method," which has underscored all the subsequent logical descriptions of the many and varied congenital cardiac lesions. These are now unified in the International Paediatric and Congenital Cardiac Code (IPCCC).[12,13] The steps involved in producing the code, now enshrined in the eleventh iteration of the International Classification of Disease, began at the World Congress held appropriately in Toronto in 2001. It was at that meeting that rapprochement to resolve the transatlantic warfare was initiated (Figure 2). Although we do not yet quite all speak the same language, at least now we are able to agree on the lesions we are describing by identifying the various phenotypes. And the advancement of clinical imaging is such that we can now

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demonstrate the salient anatomy with as much, if not more, detail as the anatomist who holds the heart in his or her hands. None of this would have proved possible without the initial concepts developed and matured by Richard Van Praagh. As Shakespeare wrote of Julius Caesar, "He doth bestride the narrow World like a colossus." And paediatric cardiac morphology, at the time, was a very narrow world. We are all indebted to his intellect and to his huge contributions. He will be sorely missed, but we will still be able to build on his firm foundations.

**Financial support.** This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

**Competing interests.** None.

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https://doi.org/10.1017/S1047951125110214 Published online by Cambridge University Press