



Joseph K. Perloff, MD: Father of Adult Congenital Heart Disease



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When a physician combines a keen intellect, insatiable curiosity, ambition, and a masterful talent for communication, it is only a matter of time before he or she is capable of rising to international prominence.

In this case the attributes describe Joseph K. Perloff, MD, author of a classic textbook and the internationally acclaimed physician who is considered the father of adult congenital heart disease.

Born in New Orleans in 1924, Dr Perloff served in the Pacific Theater during World War II. He continued his studies on returning home, receiving his medical degree from Louisiana State University School of Medicine in 1951. Following an internship and residency in medicine and pathology at Mt. Sinai Hospital in New York, he spent a formative year with Dr Paul Wood at the Institute of Cardiology in London as a Fulbright Fellow. Working under Dr Wood was an inspirational experience for the young physician that helped nurture a growing interest in cardiovascular physical examination and congenital heart disease.

When he returned to the United States and Georgetown University in Washington, DC, he rapidly rose to the rank of Professor. The keen intellect, insatiable curiosity, ambition, and a masterful talent for literary communication were key ingredients for his rapid rise to international prominence. He subsequently accepted a position at the University of Pennsylvania as Chief of Cardiology and Professor of Medicine and Pediatrics.

Dr Perloff moved to UCLA in 1977 as the Streisand/American Heart Association Professor of Medicine and Pediatrics and quickly perceived the need for an adult congenital heart disease facility. Increasing numbers of infants and children with congenital heart disease were reaching adulthood, and few cardiologists were equipped to deal with this new and complex patient population. The UCLA Adult Congenital Heart Disease Center was soon

established to provide expert patient care, physician education, and clinical and basic research. The Center rapidly gained national and international recognition, culminating in a major endowment from The Ahmanson Foundation. The Center was the first, and remains one of the largest, of such facilities in the United States.

Pursuing a long-standing interest in medical publishing, Dr Perloff wrote the classic textbook *The Clinical Recognition of Congenital Heart Disease*, which has undergone five editions. He is also the author of the widely read textbooks *Congenital Heart Disease in Adults* and *Physical Examination of the Heart and Circulation*, and of more than 400 articles in leading journals. He is on the editorial boards of *Circulation*, the *American Journal of Cardiology*, the *Journal of the American College of Cardiology*, and the *American Heart Journal*.

As the founder of a new medical specialty, Dr Perloff has enjoyed worldwide acclaim as a visiting professor and lecturer. He is the recipient of numerous national and international honors, including the American Heart Association Great Teacher Award; the United States National Physician of the Year Award; the Commemorative Medal and honorary degree in celebration of the 650th anniversary of Charles University in Prague; the Mellinkoff Award, the highest faculty honor conferred by the UCLA School of Medicine; the European/American Symposium on Congenital Heart Disease in Santorini, Greece, in honor of Professor Perloff; and most recently, the UCLA Faculty Award of Extraordinary Merit in recognition of contributions of unusually great benefit to the UCLA School of Medicine through major advances in the biomedical sciences.

On a more personal note, Dr Perloff's bedside teaching, books, lectures, and manuscripts have been instrumental in kindling and nurturing my interest in adult congenital heart disease. From personal communication with many of his other trainees, spanning three generations, I can attest to the overwhelmingly positive impression he has made. He is a masterful teacher who expects the best of himself and his students. He does not suffer fools kindly and will not tolerate shoddy or incomplete presentations.

Students willing and able to meet his stringent criteria are immeasurably rewarded by a wealth of knowledge, accumulated and meticulously sorted over nearly six decades of practice. In co-editing the third

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PH Roundtable

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Dr Gatzoulis: Because of their age and the very small number of lung transplantations taking place in the UK. I think on the organ side, perhaps you are right. This chronic cyanosis may be a good thing with the right-to-left shunting, but it is a multiorgan disease. I think, again, that any opportunity to improve organ perfusion and tissue oxygen delivery is bound to have some effect on organ function, whether it is renal or the brain, and if you can address that early in the course of the disease, and reverse the shunt, you may be able to repair the lung fields, but not many patients are suitable for this route.

Dr Krasuski: My experience has been that Eisenmenger patients can live for quite a while, and obviously it is hard to predict who is going to live and who is going to die. Unfortunately lung transplantation may not be a good exchange. Giving them a lung transplant may not give them a better survival, in fact survival could be worse, though their quality of life may improve somewhat. Timing of transplantation is a pretty tough decision.

Dr Barst: This group of patients is more difficult to compare to patients who have iPAH. But, when I discuss transplantation with patients, if they answer yes when I ask if they would ever consider transplantation, I recommend active listing when they have a poor quality of life. To me, as long as patients are able to live with their limitations and enjoy what they have in life, even if they seem sicker than other patients, I recommend they continue to do that because we don't know if they will be better or worse off after a transplant, and transplantation is a one-way street, not a panacea. Thus, overall, the timing of transplantation often be-

comes subjective and related to quality of life and to whether the patient wants it or not. I know that's not a good answer.

Dr Krasuski: Actually that is a great answer. I feel the same way.

Dr Barst: To summarize, although there remains no cure for patients with Eisenmenger syndrome, appropriate management has decreased morbidity and mortality and improved overall quality of life. With tertiary care and counseling on risks such as pregnancy, surgery, pulmonary infections, exposure to high altitude, extreme exercise, and psychological stress, patients with Eisenmenger syndrome may enjoy a better quality of life with increased survival. Current practice is to avoid interventions that may destabilize the delicately balanced physiology between the systemic and pulmonary circulations in these patients. In most cases, treatment has focused on symptomatic patients and has been directed at avoiding or ameliorating the complications associated with chronic hypoxia, hematologic abnormalities, pulmonary infection, and congestive heart failure. Prostanoids and lung or heart/lung transplantation have been shown to be effective in improving functional class and pulmonary hemodynamics. Selection criteria, however, remain problematic and the procedures are both invasive and associated with significant complications. More selective pulmonary vasodilators with antiproliferative effects hold promise in leading to improvement and better prognosis by altering the natural history of PAH associated with CHD (with or without Eisenmenger syndrome). Further investigation is needed in this patient population as extrapolation from various other forms of PAH may in fact not be applicable to PAH associated with CHD. ■

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edition of *Congenital Heart Disease in Adults* with John Child and Joseph Perloff, I came to appreciate other aspects of this great man: his friendship, his consideration for the creativity and ideas of younger colleagues, and his unwavering focus and determination to complete a given task. Despite a lifetime of

accomplishment, Joseph Perloff is not the type of man to sit back and enjoy his numerous accolades; he remains a highly productive researcher, lecturer, and writer. He is a truly inspirational individual. ■

— Jamil Aboulhosn, MD