Pediatric Cardiology: A Brief Historical Perspective

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SUMMARY. Pediatric cardiology's past covers 50 centuries. Most if its accomplishments have occurred in the last 50 years. In a limited sense, it is a story of four questions about the heart: How is it formed? How is it malformed? How can it be studied? How can it be fixed? Although not all the answers have been provided, we have come close to doing so in the first three-quarters of the 20th century, and will doubtless get even closer in the final quarter. For pediatric cardiologists and their surgeon colleagues will persist until Puck's prayer is answered:

And the blots of Nature's hand
Shall not in their issue stand.—(Shakespeare: A Midsummer Night's Dream, V,2)

Fallot [26], in his now immortal paper, said: "Nothing which touches on the solution of a clinical problem . . . should be considered devoid of interest or without importance."

A major by-product of a study of history is humility. Man invented neither illness nor its treatment. A dinosaur unearthed in Wyoming was found to have a bony tumor in a caudal vertebra. A hominid, Pithecanthropus erectus, extinct for over 400,000 years, had an exostosis of his femur. Tranquilizers and sympathomimetics were known to the ancients—Indian doctors used rauwolfia extracts, and the ancient Chinese used ephedrine. Hau-Tu, nearly 2000 years ago, narcotized his patients before operating upon them, and performed laparotomy and splenectomy. Two thousand years before Jenner, the Chinese practiced smallpox inoculation by nasal insufflation of dried crusts from a smallpox victim. The ancient Chinese also fed sheep thyroid glands to cretins. There is a tradition of bone grafting among the Arabs of North Africa, which is still being practiced, using bones from a freshly killed sheep or dog, with apparently successful results. Celsus performed tonsillectomy by enucleation over two millennia ago—a technique which has replaced the guillotine operation. Lex Caesarea (caedere = to cut), promulgated in 715 BC, declared that in the event of maternal death during pregnancy, the unborn child should be removed through an abdominal incision in the hope that it might be viable. That is how cesarean section got its name. Whether or not Julius Caesar himself was delivered by this method 600 years later, the Hindus, as well as the Romans, practiced cesarean section. Cheyne-Stokes respiration should really be called Hippocratic respiration.

These are but a few illustrations among many that could be invoked. However, they are sufficient to suggest that we exercise considerable caution before labeling any of our discoveries as new.
The study of history may lead to current application of ancient techniques, such as happened in the rediscovery of the Rauwolfia compounds. Of course, not all ancient methods were beneficial. A close inspection of standard practices of the past may serve to shatter the shibboleth of such once-revered practices as bleeding but should also caution us to examine what may well be current shibboleths, before they are scorned by our successors. Thus the study of history would be useful if only to teach us the dictum of Santayana: "Those who cannot remember the past are condemned to repeat it" (Life of Reason).

To maintain reasonable brevity, this look at the history of pediatric cardiology will be directed primarily at congenital heart disease and will focus upon the following areas: (1) the fetal circulation; (2) specific lesions; (3) cardiac catheterization; and (4) cardiovascular surgery.

The Fetal Circulation

The earliest observations on the fetal circulation are generally attributed to Aristotle [2]. He is credited by Harvey [42], with having observed pulsation in the chick embryo. Aristotle is often credited also with the first description of the ductus arteriosus. A mistranslation has persisted for two millenia due to the substitution, in an early manuscript, of arterias for arterias (aorta for trachea). Thus, what had been translated as depicting the pulmonary artery going to the bifurcation of the aorta was really a description of the superior vena cava ascending towards the bifurcation of the trachea. Since Aristotle studied only adult animals, he may have seen the ligamentum arteriosum, but probably never saw a ductus. However, Galen [33] clearly understood the ductus, and the foramen ovale:

Nature is neither lazy nor devoid of foresight. Having given the matter thought, she knew in advance that the lung of the fetus, a lung still contained in the uterus and in the process of formation and spared continual motion, does not require the same arrangements of a perfected lung endowed with motion. She has, therefore, anastomosed the pulmonary artery with the aorta, and the left and right atria... There is a certain membrane in the right atrium, connecting, in the fetus, the right atrium with the left atrium, whose appearance is rather like that of a little lid. It is easily deflected toward the pulmonary artery, ... and thereby the blood of the right atrium is prevented from flowing into the lungs. This membranous protrusion is thickened and grows together, sometimes on the first day after birth, sometimes after several days, when at length its whole body hangs down in such a way into the cavity of the vessel that it completely occludes it and it cannot be split asunder. There is also a similar projection of membrane at the mouth of the azygos vein and often at those of several other large vessels such as the jugulars, brachials and cranial veins and the trunk of the vena cava as it leaves the liver. The uses of these are the same as that of the membranes closing the mouths of the vessels of the heart.

Fallopius [25], who was usually credited with the first description of the ductus arteriosus, added nothing to Galen's description. The term ductus of Botallo persists to this day. Is Botallo to be blamed that, in investigating the fetal route from the right to the left side of the heart in calves [10], he used the term ductus to describe the channel connecting the two atriia formed by the valve of the foramen ovale? Botallo innocently scattered a seed in 1564; it was watered by Folinus [29], who reprinted Botallo's short note; and in 1660, it reappeared, thoroughly fertilized, in Van Horne's Observatio Anatomica III [80]. Van Horne annotated Botallo's text and inserted a plate that pictured the foramen ovale as described by Botallo, but also added a drawing of his own of the ductus arteriosus. Final fruition was at the hands of the Anatomical Nomenclature Commission at Basel, which harvested all under the term ductus arteriosus Botalli. Sylvius [78] describes Galen's concern with nomenclature: "How Galen had often wished it were possible to teach a thing without the use of names, for the names themselves are but the shadow of reality." But the pervasiveness of eponymity have persisted and will doubtless continue.

William Harvey [42] added little to Galen, although he did write about the fetal circulation in De generatione, a late-in-life writing that has been said to be aptly titled, "reflecting the last infirmity of a noble mind." The main criticism of the section on the fetal circulation is that the observations were entirely anatomic rather than physiologic. This same criticism can be leveled at almost all subsequent investigators until the modern era of physiologic investigation began in the 20th century. A striking exception was the work of the remarkable Oxford group who, only 40 years after Harvey, in the single decade between 1660 and 1670, identified the contribution of the lung to gas exchange, and labeled the placenta, a "uterine lung." In 1660, Robert Boyle [11] showed that part of the air is essential to life; Hooke [44] postulated that blood changes from dark to red on passing through the lungs because of its mixture with air, and Lower [54] verified Hooke's hypothesis experimentally.

For two and one-half centuries, understanding of the fetal circulation was mired in protracted arguments regarding the distribution of inferior vena cava and superior vena cava flow across the tricuspid valve or the foramen ovale, the contribution of the valve flap in the right atrium to this flow, and whether there was significant pulmonary blood