

Title: John Hunter and the Earliest Description of Three Congenital Cardiac Conditions

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Medical knowledge underwent significant advances during the 18th century's *Age of Enlightenment*. The development of reason and the scientific method coalesced to offer objective appraisal of diseases and their treatments. During this time a handful of luminaries stood out to lead the scholarship of medicine into a new era. John Hunter (1728-93) was one such individual who spearheaded the application of science to surgery. He was raised in the West Central Lowlands of Scotland as an aspiring naturalist during the *Scottish Enlightenment*, but also gained experience as an apprentice cabinet-maker. At that time this profession offered an understanding of cutting-edge design, manual craft dexterity and innovation exemplified by the works of Thomas Chippendale and other design masters of the Georgian era. Finally John was fortunate enough to receive a formal grounding in anatomy and surgery through his elder brother William Hunter following a *sink or swim test* with an arm dissection. William who remains to be considered as one of the greatest obstetricians and teachers of anatomy trained his brother over 11 years with further clinical exposure at Chelsea Hospital with William Cheselden and at St. Bartholomew's Hospital with Sir Percivall Pott.

John Hunter's multifaceted educational background likely contributed to his keen eye for comparative anatomy and disease diagnosis. He remains famous for being the first individual to describe a vast range of pathologies, treatments and anatomical discoveries.¹ These included the a deeper understanding of vascular anatomy in addition to the surgical management of popliteal artery aneurysms, studies in obstetrics and venereal disease and the identification of anatomical structures such as the Gubernaculum Testis Hunteri.² He is also acknowledged for his contribution to adult cardiology,³ being the first to describe cardiac ventricular aneurysms in 1757⁴ and pronounced to be "a gift from heaven" by the famed cardiac surgeon Henry Souttar in his 1949 Hunterian Oration.^{5, 6} Despite these acclaims, John Hunter's contribution to the discovery of several congenital cardiac conditions has been largely understated with a lack of publications supporting his role in this field. The purpose of my essay is outline for the first time John Hunter's original findings in congenital cardiac disease that have not yet been described.

Double Superior Vena Cava

By the age of 48, Hunter had amassed such surgical fame and reputation that he was appointed as surgeon to King George III in 1776. This role offered Hunter the chance to treat members of the royal household, but it also gave him the opportunity to gain access to the king's menagerie. As a keen naturalist, the king's private collection of animals which included elephants was of great interest to him. Consequently, when the captive elephants died, the king and his wife Queen Charlotte donated the carcasses of two elephants to Hunter,⁷ who subsequently dissected and studied this animal. During this dissection, Hunter was the first to identify the two pre-caval veins in the elephantine circulation.⁸ These veins represent two upper extremity channels that unify in man to represent the single superior vena cava that enters the right atrium.

Twelve years before his exposure to the king's elephants and their comparative cardiac anatomy, John Hunter was the first to describe the condition of Double Superior Vena Cava in humans as an embryological anomaly. In his case books, he describes a case from 1764 where a ten-month-old

child died from convulsions. This was the fourth in the family who had died in this manner and on autopsy Hunter described:

“The Thoracic viscera were likewise extremely sound but a remarkable circumstance of the superior vena cava, viz there were two of them: one as usual, made up of the right Subclavian and right jugular; but the other was made up of the left subclavian and left jugular; and instead of passing across the beginnings of the Carotids to join those of the right side, it passed down the left of the Heart, and received the left azygos vein, and crossed the left Ventricle, and then passed to the right, on the posterior part of the Heart, to the right auricle [and] there entered.”⁹

The persistence of the left superior vena cava (LSVC) is the most common congenital variation of thoracic venous drainage occurring in approximately 1.5%–10% of all children with congenital cardiac anomalies, although it is uncommon (0.3-0.5%) in the general population, where 85% occur in conjunction with a right superior vena cava (as in this case).¹⁰ This condition was previously thought to be first described in 1850¹¹ by John Marshall, and thus my identification of its description by John Hunter pre-dates it by approximately 80 years .

Isolation of the Left Subclavian Artery

Isolation of the left subclavian artery is a rare embryological anomaly of the aortic arch where the left subclavian artery arises from the homolateral pulmonary artery by way of a ductus arteriosus instead of coming off the aorta.¹² It typically presents with systemic cyanosis, poor left upper arm perfusion and ischaemia and cardiac failure. It is usually associated with high mortality in early life although some case reports do identify the presence of this condition in adolescence.^{13, 14} John Hunter described a case with these classical features:

“Miss Williams, a young lady about Sixteen years of age occasioned a palpitation of the Heart. When I saw her, the heart beat so forcibly that it could be seen at some distance. Its powers were very extensive..... The Carotids were seen pulsating above the Sternum. Just below the left breast between the 5th and 6th rib there was a strong thrill in the stroke. There was no pulsation in the wrist of the left arm, and that arm was very weak.....she would become black all over the left arm, and the nails of that hand”⁹

This is a classical description of a hyper-dynamic circulation and left arm ischaemia as a result of congenital cardiac disease and isolation of the left subclavian artery. This condition was previously thought to be first described in 1908 by Anton Ghon¹⁵ so that my identification of its description by John Hunter pre-dates it by approximately 150 years.

Bicuspid Aortic Valve

Bicuspid aortic valve (BAV) disease is the most common congenital cardiac defect affecting approximately 1-2% of the general population. It can lead to several cardiac pathologies including aortic stenosis, regurgitation, aneurysm formation, dissection and endocarditis.¹⁶ John Hunter described a case in 1764 with these visible features:

“We dissected a Man who had been remarkably strong, but was now very much emaciated...The Heart and Lungs were sound. On examining the valves of the Aorta, I found that there had been two only, instead of three; and that one of them had a kind of fraenum or cross-bar, attaching its middle

to the sides of the artery. These valves were very much diseased, became thick and stony, by which means they did not meet entirely, or by even edges. The left kidney had two Pelvises, and two Ureters, which entered the Bladder by two Orifices.”⁹

The co-occurrence of congenital aortic valve disease and double left renal pelvis suggests an underlying syndrome (such a Noonan syndrome), although the clear history of physical strength goes against this. Interestingly, the specimen from this case exists in the Hunterian Museum at The Royal College of Surgeons of England (**Figure 1**). The clear evidence of John Hunter’s account of a bicuspid aortic valve (which is described with a raphe) was previously thought to be first described in Sir James Paget¹⁷ in 1844 so that my identification of its description by John Hunter pre-dates it by approximately 80 years.



Figure 1. Preparation of a bicuspid aortic valve described and dissected by John Hunter in 1764, © The Royal College of Surgeons of England.

The discovery that John Hunter made the earliest description of three congenital cardiac conditions is a testament to his productivity, genius and sheer volume of work. Singlehandedly uncovering these rare cardiac conditions defines him as a great icon of medicine and surgery, however when considering these findings in the context of his multiple other contributions affords him the appropriate adulation and respect that he receives. For centuries countless surgeons, scientists and educationalists have considered him amongst the most talented and distinguished individuals in modern healthcare and biology. As a result, the continued study of his works can reveal numerous aspects of 18th century medicine, but can also inspire the persistent requirement for innovation and the scientific method to triumphantly achieve further accomplishments and improved health for the future of all mankind.

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