Brit. Heart J., 1964, **26**, 723.

NORMAN CHEVERS

A DESCRIPTION OF CONGENITAL ABSENCE OF PULMONARY VALVES AND SUPRA-VALVULAR AORTIC STENOSIS IN THE EIGHTEEN-FORTIES

BY

IVAN A. D'CRUZ AND ROBERT A. MILLER

From the Department of Pediatric Cardiology, Cook County Children's Hospital, Hektoen Institute for Medical Research and the Department of Pediatrics, University of Illinois College of Medicine, Chicago, Illinois, U.S.A.

Received March 2, 1964

Norman Chevers (1818–1886) was an English physician who has gained the recognition of medical historians because of his classic description of the pathophysiology of constrictive pericarditis (Chevers, 1842). We wish to draw attention to certain other "firsts" in the field of heart disease that lie to Dr. Chevers' credit, which apparently have been overlooked.

Norman Chevers was described in his obituary as "a man of fine intellectual capacity, high mental culture, profound learning and ceaseless industry" (Obituary, 1886). He studied medicine at Guy's Hospital, London. He joined the Indian Medical Service in 1848. Most of his work in India was done in Bengal; he became Principal and Professor of Medicine at the Calcutta Medical College and President of the Faculty of Medicine, University of Calcutta. He published numerous papers and monographs on tropical medicine, public health, and even medical jurisprudence. He was co-editor of the *Indian Annals of Medical Science* for almost twenty years. He rose to the position of Deputy Surgeon General, and on his retirement returned to England. He died in 1886 soon after completing his magnum opus "Commentary on the Diseases of India".

His claim to fame, however, rests chiefly on the work done during his twenties at Guy's Hospital, before leaving for India. During this short period he demonstrated an intense interest in cardiac pathology with its clinical correlations, publishing several papers on this subject. That this work was well appreciated at the time, we can assume from Thomas Peacock's reference to "a series of valuable and interesting papers by Dr. Chevers" in Peacock's preface to his famous *On Malformations of the Human Heart* (Peacock, 1858).

"A collection of facts illustrative of morbid conditions of the pulmonary artery as bearing upon the treatment of cardiac and pulmonary diseases" appeared as a series of papers in the *London Medical Gazette* (Chevers, 1846). He devoted a section of one of these to a discussion of congenital absence of the pulmonary valve (Chevers, 1846, p. 828). This is an entity that has attained recognition only in the past decade since the report of Lavenne *et al.* (1954). Earlier isolated reports (Kurtz, Sprague, and White, 1927; Royer and Wilson, 1908) did not emphasize the significance of the absent valve and received little attention. None of the publications on this subject mention any reference earlier than the twentieth century. Chevers discussed two cases, one described by Favell (1842) with associated tricuspid atresia, the other reported by Crampton (1830) with a ventricular septal defect and infundibular pulmonary stenosis. The description of the heart of the latter case, as summarized by Chevers, includes all the essential features of the common form of this anomaly, tetralogy of Fallot with absent pulmonary valve (Miller, Lev, and Paul, 1962): "Attached to the right ventricle there was found a supernumerary cavity which communicated with the right ventricle by an opening large enough to admit the little finger, and formed under the columnæ carnæ of the ventricle. The pulmonary artery was perfectly devoid of valves; its lining

membrane, at the usual situation of the valves, appeared a little puckered; at this spot the artery was contracted, but above it was considerably dilated. The septum of the ventricles was imperfect, and the foramen ovale patent." There is little doubt that Chevers fully realized the important role played by the absent pulmonary valve and consequent pulmonary regurgitation. Thus, in commenting on the infundibular chamber of the right ventricle, which was present in both cases, he wrote: "It is probable that this arrangement of parts in some degree compensated for the absence of a valvular apparatus, by preventing the refluent blood from being immediately thrown back into the ventricle from which it had been propelled." Chevers was apparently aware of the chief diagnostic sign of pulmonary regurgitation (the diastolic murmur) and expressed regret, "it has not been stated whether the bruit which was heard in both of these cases was synchronous with the diastole or the systole of the heart."

His account of a case of supravalvular stenosis of the pulmonary trunk (the patient of a Dr. J. Marshall) is, to our knowledge, the earliest description of such a lesion and has been overlooked by recent authors writing on this subject. He described the morbid anatomy thus: "About an inch above the valves the artery was contracted so as to form a stricture, hardened by ossific deposit, and about the diameter of a goose-quill. The coats of the artery above the stricture were very thin, and formed a sort of sac." This interesting case also had a ventricular septal defect and abnormal "shrivelled" pulmonic valve cusps.

In the same series of papers on morbid conditions of the pulmonary artery one encounters passages that reveal Chevers' remarkably advanced views on the altered physiology of cyanotic heart disease. He clearly understood and discussed at length the compensatory role of bronchial arteries in pulmonary atresia and severe tetralogy of Fallot. He postulated the part played by spontaneous closure of the foramen ovale and ductus arteriosus in the onset and aggravation of cyanosis in cyanotic anomalies, basing this upon a critical analysis of the series reviewed by him.

The earliest reference to supravalvular aortic stenosis mentioned in publications on that subject is that of Archer (1878). That type of supravalvular aortic stenosis which is associated with a typical facies, mental deficiency, and a peculiar deposition of mucinous tissue in the sub-intimal region of the ascending aorta, has attracted considerable attention of late. This particular type of aortic lesion has been recognized relatively recently. The following extract from Chevers' paper "Observations on the diseases of the orifice and valves of the aorta" (1842) leaves no doubt that Chevers was familiar with the morbid anatomy of this entity, "the portion of the artery immediately above the valves is occasionally found surrounded by a complete raised zone of tough semi-cartilaginous deposit in the subserous tissue: this may occur without being attended with the slightest apparent disease of the upper part of the vessel. Where, as is sometimes the case, this deposit receives ossific matter, it forms a permanent obstruction to the canal; but where this fails to occur, I suspect that, after a time, dilatation of the upper part of the orifice is gradually produced —the presence of the deposit appearing to cause relaxation and weakening of that part of the contractile tunic upon which it is placed."

"Observations on the structure, functions, and diseases of the coronary arteries of the heart" appeared in 1843 (Chevers, 1843). Referring to atheromatous changes in the coronary arteries, Chevers remarked, "the coincidence of this state of the coronary arteries with the symptoms of angina has long been observed." The precise relation between changes in the coronary arteries and the "fatty" or "atrophic" myocardial changes did not seem clear to Chevers, and indeed remained obscure until the end of the nineteenth century. However, he is certainly far ahead of his times in the following account of what appears to be coronary heart disease with diminished cardiac reserve and possibly attacks of "coronary insufficiency" or mild infarction. Few would disagree with his principles of therapy. "It is by no means unusual to find elderly persons of obese habits of body complaining of violent palpitation, with sensations of impending suffocation, after any sudden exertion or emotion, the application of cold to the surface of the body, or, in fact, any action which tends to determine an unusual supply of blood to the heart. In these persons, the pulse is usually weak, while percussion and auscultation shew that their hearts, although large, act

feebly; the sounds being indistinct, but free from irregularity or other abnormal character. This train of symptoms is probably often dependent upon an advanced degree of that condition of the heart which I have last described; and I have little doubt that the difficulty of breathing, which nearly all extremely corpulent persons experience upon unwonted exertion, is mainly attributable to less degrees of the same changes. Acting upon this course of reasoning, I have found that the adoption of a plan of treatment calculated at once to procure absorption of a portion of the superfluous fat of these patients, and to diminish the quantity of their circulating fluids, has been followed by an acquisition of increased cardiac power, as evidenced by a stronger pulse and an entire cessation of the suffocative attacks during very long intervals."

The tremendous flood of publications on various aspects of congenital heart disease that has appeared over the last two decades has tended to convey the impression that almost all the significant advances in this field have been of recent date. It is therefore worth pointing out that intelligent and painstaking observations, such as those made by Chevers, long preceded and foreshadowed such "advances".

References

Archer, R. S. (1878). Note on a congenital band stretching across the origin of the aorta. *Dublin J. med. Sci.*, **65**, 405.

Chevers, N. (1842). Observations on the disease of the orifice and valves of the aorta. *Guy's Hosp. Rep.*, **7**, 387.

——— (1843). Observations on the structure, functions, and diseases, of the coronary arteries of the heart. *Guy's Hosp. Rep.*, **8** (2 ser., **1**), 103.

——— (1846). A collection of facts illustrative of the morbid conditions of the pulmonary artery, as bearing upon the treatment of cardiac and pulmonary diseases. *London Medical Gazette*, **38**, 189, 276, 369, 452, 699, 744, 828, 961, 1087.

Crampton, J. D. (1830). Case of an anomalous state of the heart. *Dublin med. Trans*, n.s., **1**, 34.

Favell (1842). Malformation of heart. *Prov. Med. Surg. J.*, **3**, 440.

Kurtz, C. M., Sprague, H. B., and White, P. D. (1927). Congenital heart disease. *Amer. Heart J.*, **3**, 77.

Lavenne, F., Tyberghein, J., Brasseur, L., and Meersseman, F. (1954). Complexe d'Eisenmenger avec insuffisance pulmonaire par absence de valvules. *Acta cardiol. (Brux.)*, **9**, 249.

Miller, R. A., Lev, M., and Paul, M. H. (1962). Congenital absence of the pulmonary valve. The clinical syndrome of tetralogy of Fallot with pulmonary regurgitation. *Circulation*, **26**, 266.

Obituary (1886). Norman Chevers. *Brit. med. J.*, **2**, 1245.

Peacock, T. B. (1858). *On Malformations of the Human Heart*. Churchill, London.

Royer, B. F., and Wilson, J. D. (1908). Incomplete heterotaxy, with unusual heart malformations. Case report. *Arch. Pediat.*, **25**, 881.