Pioneers in cardiology: Dr Guy Fontaine

Guy Fontaine and arrhythmogenic right ventricular dysplasia

Cardiomyopathies are the Focus of this EHJ issue. Guy Fontaine an electrical engineer, then a physician, played a pivotal role in the discovery of ARVD.

The curious and very disturbing ‘sudden death’ of hitherto apparently healthy young athletes with no prior evidence of cardiac problems has initiated much conjecture and research, certainly since the middle of the 20th century. One of the pioneers in this field of cardiology is the renowned French cardiologist Dr Guy Fontaine MD, PhD, HDR.

Fontaine was born in Corbeil Essonnes in France. He attended secondary school at the Lycee Montesquieu in Bordeaux before moving to Paris. In addition to his medical degree, he had also trained in electrical engineering which was of great benefit in his pioneering work in electrocardiography and electrophysiology. The beginning of his medical career in the 1960s corresponded with a rapid development in medical electronics; Fontaine in fact designed and built different electronic devices for the treatment of patients presenting with electrical disorders including atrial fibrillation. He also used his electrical engineering knowledge for the advancement of pacemaker technology. For his medical thesis, he presented one device to study the cardiac pacening threshold of patients awaiting pacemaker implants. He was awarded his doctoral degree in 1966 for an excellent thesis entitled ‘Contributions to Electrical Stimulation of the Human Heart’. In 1967, Fontaine was the first physician in Paris to implant permanent pacemakers using intracardiac leads; his expertise in this field led to the referral of many patients with difficult arrhythmias from both Europe and further afield. In 1968, he joined the cardiac department at the Hôpital de la Salpêtrière in Paris led by Professor Jean Facquet.

Rather than accept any of the several offers of academic positions he received, Fontaine continued to pursue his deep interest in research into cardiac pacing. Influenced by his mentor Professor J.J. Welti and after witnessing multiple autopsies displaying ‘hearts too good to die’, Fontaine felt certain that it would be possible to eradicate ‘sudden death’ linked to atrioventricular conduction disturbances by his new approaches to solve relevant technical and biotechnical problems.

The next major topic of interest for Fontaine in 1975 was tachycardias and the use of pacemakers to treat them. He observed many episodes of chronic ventricular tachycardia (VT) preceded by extrasystoles and these appeared to be inducing their own repetition: this led him to use a properly timed stimulation of the ventricle to shorten the recovery pause after the extrasystole in order to prevent VT. Later, this concept was used to obtain the same result through stimulation of the atrium rather than the ventricle and was achieved for permanent pacing with the incorporation of a resistor in the ventricular lead connected to the normal atrial lead and on to the output of the first single-chamber digital pacemaker (Vitatron DPG-1). This design which usefully employed Fontaine’s knowledge and experience of electricity and biophysical parameters was seen as an intellectual ‘tour de force’.

In 1976, Fontaine co-authored and published, with his mentors and colleagues Professors Grosgogeat and Welti, the book ‘The Essentials of Cardiac Pacing’. At that time, he was also fascinated by the first attempts at surgical ablation of the accessory bypass tract in the Wolff–Parkinson–White (WPW) syndrome which was successfully performed at Duke University (USA) in 1968. In 1971, after 6 months of relevant research in the Christian Cabrol surgical laboratory in Paris with his colleague Dr Guiraudon, he developed the technique of epicardial mapping and they performed the first successful European surgical ablation of an accessory pathway in WPW syndrome. He further refined the technique of mapping by designing amplifiers with summation, rectification, and filtering properties, for the simultaneous recording of signals from three electrodes located at the distal end of a specially built curved probe. The purpose of this was to record potentials originating on the diaphragm surface of the ventricle,
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thus avoiding the need to raise the beating heart during surgery. A grid based on anatomical landmarks, independent of heart size, was designed and subsequently used worldwide by researchers.

After perfecting the epicardial mapping technique in animal laboratory research, the first European patient was operated on in 1971 and the report published in 1973. It was then considered possible to use the epicardial mapping in other life threatening arrhythmias, i.e. VT, and four patients with VT were successfully treated before the first arrhythmogenic right ventricular (RV) dysplasia patient; a 65-year-old male who had suffered for some 10 years from recurrent VT unrelated to coronary artery disease. The ECG suggested this patient's tachycardia originated in the RV—an interesting finding since in the previous four patients, the site of origin was the left ventricle.

The operation took place on 30 October 1973, and on exposure, the surgeon immediately noted moderate dilation of the RV which was hypokinetic and covered by an unusually significant layer of fat. Mapping, which took around 20 min, confirmed the 'site of origin' to be in the mid-RV free wall. A 'simple ventriculotomy' was performed along the anterior border of the RV which showed an apparently normal RV cavity. Given the poverty of the muscular wall essentially replaced by fat, an RV biopsy had been difficult.

The histology of the fragment showed no signs of inflammation. Closure of the ventricle and withdrawal of extracorporeal circulation followed by burst stimulation did not result in inducible tachycardia: a very satisfactory result and without the need for anti-arrhythmic drugs. Fontaine's team was convinced that a surgical treatment of resistant VT had been discovered. The patient died of cardiac failure aged 86 years but without further episodes of VT. The results were presented in 1975 at an international meeting in Amsterdam.1 Another international meeting in Liege, Belgium, in 1976, Fontaine found it necessary to propose a new name for this recently discovered clinical entity.2 'Arrhythmogenic' was an obvious choice as was 'right ventricular' since both described salient features of original referrals. Lastly, 'dysplasia' was chosen to cover what appeared to be a developmental problem. Given the young age of patients, it was thought that the disease started early in development, or as we now know, in embryo—and thus the birth of 'Arrhythmogenic Right Ventricular Dysplasia'.2–4

Fontaine has written 864 scientific articles and at least 195 book chapters, many published in English. He became expert in the pathology of the RV to explain the natural history of the disease.4 He has received numerous honours and awards and the French 'Legion d'Honneur' is pending. He has been invited to lecture worldwide. He is rightly considered as the 'epitome of the international scientist'. His interests and hobbies include classic music and the new unique style of 2300 'Calligraphic Paintings and Drawings'.

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References