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Arrhythmogenic right ventricular dysplasia

Sir,—The three consecutive articles on arrhythmogenic right ventricular dysplasia (ARVD) published in the February issue of the *British Heart Journal* demonstrate the increasing interest in this clinical entity.[1–3]

Gerlis *et al* conclude that there is a clear distinction between ARVD and Uhl's anomaly. This accords with our limited experience of these two conditions.[4] The confusion probably arose because Uhl's original description was based on only one case.[4] Because Uhl's anomaly was regarded as the most striking example of a condition affecting the right ventricular musculature any disorder of the right ventricular myocardium was viewed as a form of Uhl's anomaly.[5] Later it was realised that infiltration of the myocardium of the right ventricular free wall by fatty tissue with few remaining myocardial fibres did not fit Uhl's original description.[6]

There has been some confusion about the so-called "partial" and "complete" forms of the disease. The term "partial" was originally used by Sugiura *et al* who described a heart obtained at necropsy that had "apposition of the epicardium on the endocardium", which appeared as a small oval window (12 mm wide and 15 mm long) on the free wall of the right ventricle. This was an incidental finding because the patient had no cardiac symptoms.[7] Another striking example of Uhl's anomaly was seen at necropsy in a patient with a normal cardiac examination before death. In this patient, only the anterior free wall of the right ventricle was paper-thin and translucent.[8] Therefore, the term "partial" should be restricted to patients in whom an area of the heart is completely devoid of muscle and should not be applied when there is only a "partial replacement" of the right ventricular musculature by fatty tissue in the free wall of the right ventricle. When some cardiomyocytes remain between the epicardium and endocardium in the most affected area, Uhl's anomaly should not be diagnosed. Dr Lino Rossi, a pathologist of great experience, told me that in 40 years he had never seen a typical case of Uhl's anomaly.

We saw our first adult case of Uhl's anomaly in 1974.[9] The patient was referred to us because of our special interest in the surgical treatment of ventricular tachycardia. The right ventricle was monstrously dilated and the wall of the right venticle was so thin that at surgery blood could be seen flowing inside the right ventricular cavity. This case was the first in which the reentrant pathway of ventricular tachycardia was mapped on the anterior aspect of the infundibulum where some fibres remained, making a two dimensional structure.[10] We recorded late potentials from this area 260 ms after the onset of the QRS complex.[11] This patient died of atrial fibrillation and uncontrollable heart failure after operation. The biopsy specimen taken at operation showed "abrupt interruption of myocardium" with total absence of ventricular muscle and apposition of epicardium against endocardium in most of the free wall of the right ventricle.

Our second case was a patient who died of pulmonary emboli before surgery for ventricular tachycardia could be done. The cardiac pathology in this case has been reported elsewhere.[10 12 13] Histological examination showed absence of myocardial muscle and again the abrupt interruption of the myocardium. These two cases were reviewed by our cardiac pathologist Dr Fabrice Fontaliran, who confirmed that these patients had Uhl's anomaly. Therefore, we think that the cases reported by Vedel *et al*[10] in which the pathology was not fully described were probably two adult cases of Uhl's anomaly.

Miani *et al* discussed the most difficult aspects of the differential diagnosis.[2] They presented two families that illustrated the role of left ventricular involvement in ARVD. Fatty tissue can be a normal component of the right ventricle (but not the left), and the common feature of the pathological material available from the two families was fibrosis of both ventricles.[14] In ARVD a moderate amount of fibrous tissue is generally found around the surviving myocardial fibres that are embedded in the fatty tissue. This was not the pattern of fibrosis shown in the cases of both families. The description of the pathological examination in these cases (especially the first family) is more compatible with the fibrous form of idiopathic dilated cardiomyopathy.

In addition, the progression of the disease is quite different in these two families. In the first family, the involvement of the left ventricle was the most salient feature in all the cases, and ventricular tachycardia, probably originating in the right heart, was seen only in the third case. The presence of lymphocytic and plasmocytic infiltration suggests that cardiomyopathy was the result of earlier myocarditis. The progression of the disease in the second family resembles that of ARVD.[1 2 8 9] However, we cannot exclude the possibility that the pathological findings could be the result of a healed myocarditis in which the signs of a previous infection had disappeared.

An abnormal host immune response could explain the familial cases and the histological "fibrous pattern" that was more frequently seen in the Veneto region than in our own series.[15] The cases reported by Gerlis *et al* are examples of the congenital form of dysplasia and those reported by Miani *et al* are probably examples of the acquired form of the disease. It is likely that both ARVD and Uhl's anomaly are the result of abnormal development[16 17] rather than a pathological entity caused by myocarditis alone.[18] ARVD could be a congenital anomaly with superimposed myocarditis. In either event the term dysplasia, which means abnormal development as well as a pathological structure resulting from an inflammatory process, deserves to be reinstated.[19]

McLay *et al* reported their interesting experience with treating ventricular tachycardia in patients with ARVD who did not respond to antiarrhythmic drugs.[3] Even when patients with ARVD have considerable dilatation of the right ventricle or rapid progression of right ventricular enlargement and ventricular tachycardia with multiple configurations, we still consider ablation with radiofrequency or DC energy, provided that strict protocols are followed. This approach is used alone or in combination with drug treatment at our hospital and others, and was effective in the short and long term.[20 21] We think that surgery should be performed only when there is an additional indication—such as, correction of abnormal venous return, removal of pacing leads in case of sepsis, etc.

I hoped that the long-term study of outcome in the patients who have had the operation described by McLay *et al* (disconnection of the right ventricle) will include not only patients with ARVD who have this procedure but also matched controls.

Finally, I agree that ARVD is not a rare clinical entity, as indicated by the two cases of McLay *et al*. ARVD could cause sudden death in a young adult who is symptom free or has experienced only minor cardiac symptoms before the terminal event. Because ARVD may not be uncommon it is important to screen individuals who apply for positions in which sudden illness or loss of consciousness caused by ventricular tachycardia or ventricular fibrillation would pose a considerable risk to others. They could be screened by echocardiography for several conditions, including hypertrophic cardiomyopathy and ARVD. Independently, patients could be screened for ARVD by ordinary and signal averaged electrocardiograms. The cost benefit ratio would depend upon the prevalence of ARVD in the population that is studied, and this is as yet unknown.

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SIR,—Gerlis *et al* (*Br Heart J* 1993;69: 142–50) tried to achieve a more precise definition of the entity(ies) in which the myocardium of the right ventricle is absent or scarce. This is an important aim because it could improve our understanding of the processes and have an impact on genetic counselling, early diagnosis, and treatment of the diseases. This goal is hindered by the rarity of the diseases and by the absence of a good correlation between clinical and morphological findings. Some patients that fulfil the clinical and electrophysiological criteria for the diagnosis of right ventricular dysplasia do not have fat replacing the right ventricular myocardium: on the other hand, some patients with this morphology present with congestive heart failure rather than arrhythmia or have atypical electrophysiological findings.

As well as the cases reviewed by Gerlis *et al* others are reported by American groups that use a descriptive name—partial absence of the myocardium of the right ventricle.[1] I and colleagues described such a patient in whom the right third of the septum was replaced by fat, but not the left two thirds.[2]

We reported an important morphological feature also mentioned by Fontaine *et al*[3] but not by Gerlis *et al*[2]—medial thickening and partial destruction of the elastic fibres of the intramural coronary vessels. This finding should be helpful in the characterisation of right ventricular dysplasia and also could give additional clues about its pathogenesis.

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This letter was shown to the authors, who reply as follows:

SIR,—We concur with the additional points raised by Dr Gutierrez.

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BOOK REVIEW

Organ transplantation: long-term results. Edited by Leendert C Paul and Kim Solez. (Pp 413; $145). New York: Marcel Dekker, 1992. ISBN 0-8247-8599-1.

This book brings together the long-term complications of transplantation of the various solid organ allografts. The editors are well respected experts on transplantation, as are the authors of the individual chapters. The book is very well referenced throughout. The strongest chapters are those of more general relevance to all allografts and concern immunology, general features of chronic rejection, malignancy, and skin changes.

Inevitably, in a multiauthor book there is some repetition particularly in relation to graft vascular disease. Chapters on chronic rejection changes in individual allografts are generally the least inspiring in that the information is readily obtainable elsewhere. Nevertheless it is useful to have chapters on the various grafts within one book—particularly as a good source of references.

There is a chapter on cardiac graft atherosclerosis and one on chronic heart graft rejection in the clinical setting. Both of these adequately cover the relevant topics and problem areas. It is surprising that in the chapter on heart/lung and lung transplantation the authors concentrate on obliterative bronchiolitis. Chronic graft vascular disease certainly occurs in the transplanted lung and furthermore graft vascular disease in hearts in combined heart/lung transplantation is worthy of discussion. Though obliterative bronchiolitis in these transplants at present is the principal factor limiting long-term survival, as strategies develop to curtail this, graft vascular disease both in the lung and in the heart may become more important. That occurring in the heart is particularly interesting as it seems to be less common than in orthotopic heart transplants and this provides useful information concerning possible aetiological factors—for example, quality of donor organ, organ preservation, and cytomegalovirus infection.

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BRITISH CARDIAC SOCIETY NEWSLETTER

News from Council

The Council of the British Cardiac Society at its meeting on the 7 July reviewed recommendations from the Programme Committee concerning the Annual Meeting in Torquay next year. It was decided to confine the meeting to three days from the 17 to 19 May inclusive (Tuesday to Thursday). The Annual Business Meeting and the Annual Dinner will be held on the evening of Wednesday 18 May. To facilitate the full review of abstracts, the abstract submission date has been brought forward to 1 December (from 15 December). The Young Research Workers Prize, which is attracting an increasing number of submissions, has been reviewed by Council and it has been decided that submissions for the prize will be limited to 3000 words excluding references. The closing date for submission is being brought forward to 1 November 1993. Moderated poster sessions are now to be held on each of the three days during a longer morning coffee break. This follows the successful trial session of moderated posters at Wembley.

Council has decided to broaden the membership of the Society by encouraging applications from registrars who are committed to training in cardiology. Previously, with a few exceptions, only senior registrars and consultants were considered for membership. Applications for membership are required by 1 December 1993 for consideration for enrolment next year and