Arrhythmogenic right ventricular dysplasia

Sir,—The three consecutive articles on ar-

rhymogenic 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 anom-
yaly. This accords with our limited ex-
terence of these two conditions.4 The
confusion probably arose because Uhl's or-
iginal description was based on only one

case.5 Because Uhl's anomaly was regarded as
an 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.6 Later it was realised that
infiltration of the myocardium of the right ven-
tricular free wall by fatty tissue with few remain-
ning myocardial fibres did not fit Uhl's original
description.7

There has been some confusion about the
so-called "partial" and "complete"
forms of the disease. The term "partial" was
originally used by Sugita et al who des-
cribed a heart obtained at necropsy that had
"apposition of the epicardium on the endo-
cardium", 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
inconclusive finding because the patient had
no cardiac symptoms.8 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 ven-
tricular wall was paper-thin and translucent.9
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 ventric-
lar musculature by fatty tissue in the free
wall of the right ventricle. When some car-
diomyocytes remain between the epic-
cardium 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 1974.4 The patient was referred to us
because of our special interest in the
surgical treatment of ventricular tachycar-
dia. The right ventricle was monstrously
dilated and the wall of the right ventricle was
so thin that at surgery blood could be seen
flowing inside the right ventricular cavity.
This case was the first in which the reen-
trant 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 com-
plex.11 This was typical of atrial fibril-
lation and uncontrollable heart failure after
operation. The biopsy specimen taken at
operation showed "abrupt interruption of
myocardium" which "had disappeared at the
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 embolism. Before surgery ventricu-
lar tachycardia could be done. The

cardiac pathology in this case has been
described elsewhere.10 Histological exam-
ination showed absence of myocardial
muscle and again the abrupt interruption of
the myocardium. These two cases were re-
viewed by our cardiac pathologist Dr
Fabrice Fontaliran, who confirmed that
these patients had Uhl's anomaly. There-
fore, we think that the cases reported by
Veled et al11 in which the pathology was not
fully described were probably two adult
cases of Uhl's anomaly.

Miani et al12 have described the most difficult
aspects of the differential diagnosis. They
presented two families that illustrated the
role of left ventricular involvement in
ARVD. Fatty infiltration can be a normal con-
comitant of the right ventricle (but not the
left), and the common feature of the patho-
logical material available from the two fam-
ilies was fibrosis of both ventricles.13 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 pathological examina-
tion in these cases (especially the first
family) is more compatible with the fibros-
form of idiopathic dilated cardiomyopathy.

In addition, the progress of the dis-
 ease 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,
which probably originated from the left 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 a patient who is symptom free,
also matches that of ARVD.14,15 However, we
cannot exclude the possibility that the patholo-
gical 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 his-
tological "fibrous pattern" that was more
frequently seen in the right ventricle than in
our own series.16 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 development17 rather
than a pathological entity caused by myo-
carditis alone.18 ARVD could be a myocar-
dial 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 re-
instated.19

McLay et al reported their interesting ex-
terence with treating ventricular tachy-

3 McLay JS, Norris A, Campbell RW, Kerr F. Arrhythmogenic right ventricular dysplasia: an uncommon cause of ventricular tachy-
6 Virmani R, Robinowitz M, Clark MA, McAllister HA. Sudden death and partial absence of the right ven-
8 Slama R, Leclercq JF, Counilh Ph. Parox-
tricular cardia in patients with ARVD who did not respond to antiarrhythmic drugs. Even when patients with ARVD have consider-
able dilation of the right ventricle or partial progression of right ventricular enlargement and ventricular tachycardia with multiple configur-
ations, we still consider ablation with radiofrequency or DC energy, pro-
vided that strict protocols are followed. This approach is used alone or in combi-
nation with drug treatment at our hospital and others, and was effective in the short and long term. Procurement of the right ventricle should not be performed only when there is an addi-
tional indication—such as, correction of abnormal venous return, removal of pacing leads in case of sepsis, etc.
10 I hoped that the long-term study of out-
come in the patients who have had the operation described by McLay et al (discon-
nection of the right ventricle) will include not only patients with ARVD who have this procedure but also matched controls.
11 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 has been considered healthy or has experienced only minor cardiac symptoms before the terminal event. Because ARVD may be uncommon it is important to seek individuals who apply for positions in which sudden illness or death 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. Independ-
ently, patients could be screened for ARVD by ordinary and signal averaged echocardiography. The cost:benefit ratio would depend upon the prevalence of ARVD in the population that is studied, and this is as yet unknown.

GUY FONTAINE
Hôpital Saint-Rosan
94200 Ivry, France


Sir,—Gerlis et al (Br Heart J 1993;69: 142–50) tried to achieve a more precise defini-tion of the entity(ies) in which the myocardium of the right ventricle is absent or scarce. This is an important aim because it could help in 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 condition and by the absence of a good correlation between clinical and mor-phological findings. Some patients that fulfil the clinical and electrophysiological criteria for the diagnosis of right ventricular dysplasia do not present the right ventricular myocardium: on the other hand, some patients with this morphology present with congestive heart failure rather than arrhythmias and have atypical electrophysi-o logical 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 al3 but not by Gerlis et al—mediated thickening and partial destruction of the elastic fibres of the intramural coronary vessels. This finding should be helpful in the characterization of right ventricular dysplasia and also could give additional clues about its patho-geneis.

PAULO SAMPAIO-GUTIERREZ
Department of Pathology—SM-30,
University of Washington,
559 NE Pacific Avenue,
Seattle, WA 98195,
USA


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

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

R H ANDERSON
Department of Pathology,
Royal Brompton National Heart and Lung Institute,
Dover Street,
London SW3 6LY

BOOK REVIEW


This book brings together the long-term complications of transplantation of the vari-ous 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 through-out. 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—partic-u larly 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 trans-plantation the authors concentrate on obliterator bronchiolitis. Chronic graft vascular disease certainly occurs in the transplanted lung and furthermore graft vascular disease in hearts in combined heart/lung transplan-tation is worthy of discussion. Though with the recently increased number of transplants at present this 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 trans-plants and this provides useful information concerning possible aetiological factors—for example, quality of donor organ, organ preservation, and cytomegalovirus infection.

N R B CARY

If you wish to order or require further information regarding the titles reviewed here, please e-mail or telephone the BMJ Bookshop, PO Box 295, London WC1H 9TR. Tel: 071–383 6244. Fax 071–383 6662. Books are supplied post free in the UK and for BFPO addresses. Overseas customers should add 15% for postage and packing. Payment can be made by cheque in sterling drawn on a UK bank, or by credit card (MasterCard, Visa, or American Express). Please include your card number, expiry date, and your full name. (The price and availability are occasionally subject to revision by the Publishers).

BRITISH CARDIAC SOCIETY

NEWSLETTER

News from Council
The Council of the British Cardiac Society at its meeting on the 7 July reviewed recommen-dations from the Programme Commit-tee concerning the Annual Meeting in Torquay next year. It was decided to con-fine 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 Decem-ber (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 this prize will be limited to 3000 words excluding refer-ences. 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 com-mitted 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 con-sideration for enrolment next year and