steroid contraceptive—have clear limitations. Neither is well suited to the needs of these young, backward girls. There is growing concern over the problem of pelvic infection resulting from the use of intrauterine contraception, particularly among young, nulligravid girls.[1 2] Moreover, the risk of expulsion and hence of pregnancy is significantly higher among nulligravidae in the youngest age groups.[3 4] Again, these girls are ill equipped to cope with the menstrual irregularities and cramp-like pains that may accompany the use of an intrauterine device. In Britain the Committee on Safety of Medicines has not yet given general approval to the use of long-acting progestogen injections for contraceptive purposes, reports of abnormal patterns of bleeding and of troublesome mood changes, breast discomfort, and weight gain being causes of concern.[5]

Understandably, some parents raise the question of sterilisation and their request is not unreasonable, though it does raise profound moral and ethical issues to which there are no easy answers. This was frankly acknowledged by Dr David Owen as Secretary of State for Health and Social Security when the issue was discussed in the House of Commons in June 1975.[6] In his statement to Parliament Dr Owen agreed that regrettably there were circumstances when a child under the age of 16 might best be sterilised. "What we have a duty to do," he added, "is to ensure that those decisions, when they are made, are made in a manner which will be acceptable to public opinion as a whole." In a later statement in the House[7] he referred to the case of a young Sheffield girl who had been made a ward of court by Mrs Justice Heilbron as a means of blocking a proposal to sterilise her. In the light of this case, after conferring with its expert advisers the Department of Health in October 1975 issued to all area and regional health authorities a discussion paper setting out proposals that it was hoped would result in an agreed code of practice to be followed by doctors faced with a request to sterilise a minor for non-therapeutic reasons. In effect, this document proposed that no single doctor should assume responsibility for sterilising a minor. All those able to provide information about the medical, psychological, educational, and social problems of the child should be consulted as well as the parents, without whose consent the matter could not proceed. Where doubt or argument arose the paper suggested that the case should be referred to a local, independent ethical committee with lay as well as professional members. And there the matter rests. The department has offered no firm guidance and no code of practice has been agreed.

These cases must be managed in conformity with the present law. Before any operation is undertaken informed consent is essential, and the most important aspect of any consent procedure is the duty to explain to the patient the nature and purpose of the proposed operation and to obtain fully informed consent. Persons of unsound mind cannot give a valid and fully informed consent, and if these young girls are incapable of appreciating fully the nature and consequences of a sterilising procedure then it would be unlawful to subject them to the operation. Arranging for a committee to make a decision about such an operation might look like an attempt to shed some of the responsibility on to others, whereas the legal responsibility lies clearly with the surgeon who undertakes the operation. The law at present is not permissive in regard to the sterilisation of mentally retarded minors, and any doctor approached by parents with a request for such an operation would be well advised to seek advice from his defence society. Each case will differ in some respect from the next, and all the circumstances would have to be weighed carefully before any surgeon took it on himself to challenge the law in the way that Bourne did over therapeutic abortion over 40 years ago. But the law, representing society, is likely to take a more serious view of a sterilising operation, which is in large measure an irreversible procedure.

We cannot assess accurately how many families would like such an operation carried out, for some parents, aware of the present difficulties, will not approach their doctor. Were it to be established openly that medicine and the law are prepared to give a sympathetic and understanding hearing to their problem more families might seek help. But there is no possibility of sterilising even the most seriously affected children within the framework of the law as it stands—from every viewpoint a most unsatisfactory state of affairs.

[1] United States Department of Health, Education and Welfare, Food and Drug Administration. Medical Device and Drug Advisory Committee on Obstetrics and Gynecology. *Second report on intrauterine contraceptive devices.* Washington, DC: US Government Printing Office,1978.
[2] Guillebaud J. Pelvic inflammatory disease and IUCDs. *British Journal of Family Planning* 1978;**4**:25, 29-30.
[3] Bernard RR. IUD performance patterns: a 1970 world view. *Int J Gynaecol Obstet* 1970;**8**:926-40.
[4] Guillebaud J. The safety of intrauterine devices. *Stud Fam Plann* 1979;**10**: 174-7.
[5] Savage W. The use of Depo-Provira in East London. *Fertility and Contraception* 1978;**2**:41-7.
[6] *House of Commons Official Report (Hansard)* 1975 June 25;**894**:cols 633-8.
[7] *House of Commons Official Report (Hansard)* 1975 July 21;**896**:col 248.

Arrhythmia in hypertrophic cardiomyopathy

Patients who die from hypertrophic cardiomyopathy most often do so suddenly.[1] Before the fourth decade of life symptoms tend to be mild and often respond to treatment with beta-adrenergic-blocking drugs; the patient usually becomes ill as his haemodynamic condition deteriorates and he develops atrial fibrillation and heart failure.[2] Patients who are in atrial fibrillation with grossly raised venous pressures and a low cardiac output might be expected to die suddenly, presumably from ventricular fibrillation. What is disturbing is that sudden death strikes down not only patients with advanced disease but also younger, often asymptomatic patients. In one series of 220 patients with hypertrophic cardiomyopathy followed up for a mean of six years, 27 patients died suddenly: most were younger than 40 years and were asymptomatic before death.[3]

Continuous electrocardiographic monitoring of patients outside hospital has shown that asymptomatic arrhythmia is surprisingly common in patients with hypertrophic cardiomyopathy[4]: an investigation at the Royal Postgraduate Medical School showed that 50% of such patients had serious ventricular arrhythmias and half of these had ventricular tachycardia. Similar results have been reported by other groups.[5] Treatment with beta-adrenergic-blocking drugs (mean dose of propranolol 280 mg/day) did not reduce the number of attacks of supraventricular or ventricular arrhythmias.[4] Recently the calcium antagonist verapamil has been advocated as an alternative treatment,[6 7] but in short-term studies (two months) it was not shown to reduce the number of episodes of arrhythmia in hypertrophic cardiomyopathy.[8]

The questions that need to be answered are whether arrhythmia is the cause of sudden death, and if so whether successful treatment of arrhythmia is possible.[9] A prospective study using ambulatory electrocardiographic monitoring showed that patients found to have episodes of ventricular

tachycardia were more likely to die suddenly during the follow-up period of one to four years.[10] At the very least, the evidence suggests that control of ventricular arrhythmias may help prevent sudden death. As more experience is gained the prospects seem better for the control of these arrhythmias with drugs.[8 11] Long-term, ambulatory electrocardiographic monitoring will be required in all patients with hypertrophic cardiomyopathy so that serious ventricular arrhythmias may be detected and treated early on.

[1] Maron BJ, Roberts WC, Edwards JE, McAllister HA, Foley DD, Epstein SE. Sudden death in patients with hypertrophic cardiomyopathy: characterization of 26 patients without functional limitation. *Am J Cardiol* 1978;**41**:803-10.

[2] Hardarson T, De La Calzada CS, Curiel R, Goodwin JF. Prognosis and mortality of hypertrophic obstructive cardiomyopathy. *Lancet* 1973;ii: 1462-7.

[3] McKenna WJ, Deanfield JE, Faruqui AM, Oakley CM, Goodwin JF. Prognosis and mortality in hypertrophic cardiomyopathy. *Circulation* 1979;**59,60,** suppl II:II-154.

[4] McKenna WJ, Chetty S, Oakley CM, Goodwin JF. Arrhythmia in hypertrophic cardiomyopathy: exercise and 48-hour ambulatory electrocardiographic assessment with and without beta adrenergic blocking therapy. *Am J Cardiol* 1980;**45**:1-5.

[5] Savage DD, Seides SF, Maron BJ, Myers DJ, Epstein SE. Prevalence of arrhythmias during 24-hour electrocardiographic monitoring and exercise testing in patients with obstructive and nonobstructive hypertrophic cardiomyopathy. *Circulation* 1979;**59**:866-75.

[6] Kaltenbach M, Hopf R, Kober G, Bussmann W-D, Keller M, Petersen Y. Treatment of hypertrophic obstructive cardiomyopathy with verapamil. *Br Heart J* 1979;**42**:35-42.

[7] Rosing DR, Kent KM, Maron BJ, Epstein SE. Verapamil therapy: a new approach to the pharmacologic treatment of hypertrophic cardiomyopathy. II. Effects on exercise capacity and symptomatic status. *Circulation* 1979;**60**:1208-13.

[8] McKenna WJ, Harris L, Perez G, *et al.* Hypertrophic cardiomyopathy: comparison of verapamil and amiodarone in the treatment of arrhythmias. *Br Heart J* (in press). (Abstract.)

[9] Goodwin JF, Krikler DM. Arrhythmia as a cause of sudden death in hypertrophic cardiomyopathy. *Lancet* 1976;ii:937-40.

[10] McKenna WJ, England D, Oakley CM, Goodwin JF. Detection of arrhythmia in hypertrophic cardiomyopathy: prospective study. *Am J Cardiol* (in press). (Abstract.)

[11] Canedo MI, Frank MJ, Abdulla AM. Rhythm disturbances in hypertrophic cardiomyopathy: prevalence, relation to symptoms and management. *Am J Cardiol* 1980;**45**:848-55.

Pathophysiology of Raynaud's phenomenon

Raynaud's phenomenon is episodic digital ischaemia provoked by stimuli such as cold,[1] emotion,[2] trauma,[3] hormones,[4] and drugs.[5] It is manifested by pallor of the affected digits, followed by cyanosis and then redness; these changes reflect, respectively, the underlying arterial ischaemia, venostasis, and reactive hyperaemia. In some cases the underlying episodic vasospasm is severe enough to close the digital arteries completely,[6] but the precise mechanism is still not fully understood.

Primary Raynaud's disease is found in otherwise normal healthy individuals. The anatomy of the digital arteries of these patients shows no variation from normal.[7] Raynaud's phenomenon is secondarily associated with a whole host of diseases, including occlusive arterial disease, neurovascular entrapment syndromes, and many of the connective-tissue diseases. Of these, the strongest association is with systemic sclerosis, where up to 90% of patients may have Raynaud's phenomenon. A disease closely related to systemic sclerosis and identified by the presence in the serum of an antibody to ribonuclear protein is mixed connective-tissue disease.[8] Patients with this disease may show features of systemic lupus erythematosus, dermatomyositis, or systemic sclerosis. Most have severe Raynaud's phenomenon. The aetiology of this disease is unknown, whereas in systemic sclerosis the lesions in the small vessels are well described and, indeed, the vascular tree may be seen as the main target organ.[9] What these associated diseases have in common is that their pathophysiological changes affect blood flow—through changes in pressure gradient, vessel calibre, or blood viscosity.

Apart from any structural changes that may reduce their calibre, the diameter of the digital arteries depends on the physiological mechanisms that regulate vasomotor tone. These arteries play a vital part in regulating body temperature, and blood flow through them may vary up to 200-fold.[10] Blood flow is reduced in both primary Raynaud's disease and secondary Raynaud's phenomenon under basal and "cold"-provoked conditions.[10–12]

Vasomotor tone is influenced reflexly by the sympathetic nervous system and directly by local factors including chemical mediators such as serotonin[13] and prostaglandins.[14] The adrenergic sympathetic fibres constrict the digital arteries, but since these arteries have no vasodilatory fibres supplying them reflex dilatation occurs when the sympathetic influence is removed. Raynaud himself considered that his phenomenon was due to excessive sympathetic activity; but over 120 years this theory has never been substantiated and the many treatments directed towards minimising sympathetic control of the vasomotor tone have given disappointing results. Sir Thomas Lewis[15 16] later proposed the "local-fault" theory, suggesting that the vessels themselves were abnormal. Our understanding of these "local" factors has made little progress but discovery of the prostaglandins may have helped. The vasodilatory action of the E and F classes of prostaglandins has long been recognised, but only relatively recently have thromboxane A_2 and prostacyclin (PGI_2) been identified. Thromboxane A_2, produced largely in platelets, is a potent vasoconstrictor and platelet aggregator.[17] Prostacyclin is synthesised in and released from vascular endothelium[18] and is a potent vasodilator and inhibitor of platelet aggregation.[19] The interaction of these two compounds is thought to control the laying down and clearing of platelet thrombi within the vasculature. Lewis performed some very elegant studies on the digital arteries from patients with Raynaud's disease and Raynaud's phenomenon. Those patients who had had tissue necrosis and ulceration were found to have thrombi in the small digital vessels,[7] a finding later confirmed by Pickering.[20] Most probably prostaglandin and thromboxane A_2 will prove important factors in the process.

Finally, blood viscosity may play a part in Raynaud's phenomenon. Pringle *et al*[21] and Goyle and Dormandy[22] reported that the viscosity was raised in patients with both Raynaud's disease and Raynaud's phenomenon. This increase in viscosity was more definite at 27°C than at body temperature —yet another factor impeding blood flow in the cold. Thus each of the factors that influence blood flow may be abnormal in both Raynaud's disease and Raynaud's phenomenon—though we still do not understand the roles of these abnormalities in the pathogenesis of this reversible digital ischaemia.

[1] Raynaud AGM. *De l'asphyxie locale et de la gangrène symetrique des extrémités.* Paris: Rignoux, 1862.

[2] Fox RH. Effects of cold on the extremities. *Proc R Soc Med* 1968;**61**:785-7.

[3] Loriga G. Pneumatic tools. In: *Occupation and health encyclopaedia of hygiene, pathology and social welfare.* Vol 2. London: International Labour Office, 1934.

[4] Eastcott HH. Raynaud's disease and the oral contraceptive pill. *Br Med J* 1976;ii:477.

[5] Marshall AJ, Roberts CJC, Barritt DW. Raynaud's phenomenon as side effect of beta-blockers in hypertension. *Br Med J* 1976;i:1498-9.