Echocardiographic assessment of left ventricular function in Duchenne's muscular dystrophy

M. AHMAD,¹ J. E. SANDERSON, V. DUBOWITZ, AND
K. A. HALLIDIE-SMITH

From Departments of Medicine (Clinical Cardiology) and Paediatrics (Jerry Lewis Muscle Research Centre), Royal Postgraduate Medical School, Hammersmith Hospital, Du Cane Road, London

SUMMARY Left ventricular function was assessed echocardiographically in 13 children with Duchenne's muscular dystrophy. Ejection fraction, mean velocity of circumferential fibre shortening, normalised left ventricular posterior wall velocity, maximal systolic endocardial velocity, and maximal diastolic endocardial velocity were determined manually by measuring directly from the echocardiographic tracings. The echocardiograms were then digitised and results computed in order to study the peak rates of change of dimension, the peak lengthening rates, the peak shortening rates, and the time relation between the mitral valve and left ventricular posterior wall movement in early diastole.

All the systolic phase indices were within normal limits. Though maximal diastolic endocardial velocity (11.1 ± 3.2 cm/s = mean $\pm$ SD) was reduced in patients with Duchenne's muscular dystrophy compared with normal controls (15.3 ± 3.2 cm/s), the peak rates of change of dimension and the peak lengthening rates were identical with normal subjects. The time relation between the mitral valve and left ventricular posterior wall movement was not significantly different from normal. The maintenance of this close time relation and the fact that the ventricle is filling normally, as evidenced by normal peak filling rates, suggest that the myocardial function is well preserved in children with Duchenne's muscular dystrophy. The reduced maximal diastolic endocardial velocity may be the result of reduced overall movement of the heart within the thorax.

Cardiac involvement is now a well-recognised feature of Duchenne's muscular dystrophy though heart failure is not a common cause of death. Electrocardiographic abnormalities are frequent and until recently have given the earliest indication of myocardial involvement. Since the changes in the electrocardiogram commonly suggest abnormality of the posterior left ventricular wall, a part of the heart which is well suited to study by echocardiography, the use of this non-invasive technique is particularly attractive for the study of children. Kovick *et al.* (1975) reported an echocardiographic study of patients with all forms of progressive muscular dystrophy including 3 patients with Duchenne's muscular dystrophy and we have now amplified their work in a larger series of 13 children using a simple digitising technique to analyse the echocardiogram. Left ventricular systolic as well as diastolic functions were evaluated.

Methods and subjects

Thirteen patients with classical Duchenne's muscular dystrophy attending the Muscle Clinic were studied. The diagnosis was confirmed by biochemical, electromyographic, and muscle biopsy data. All the patients were in sinus rhythm except one who had junctional rhythm. None was on any drug treatment.

Seventeen subjects matched for age without clinical evidence of heart or skeletal muscle disorder were also studied.

ECHOCARDIOGRAPHIC TECHNIQUE

The echocardiographic studies were performed with an Ekoline 20 Ultrasonoscope (transducer frequency 2.25 MHz, repetition frequency 1000 pulses per second). The output was displayed on a Cambridge Strip Chart Recorder at a paper speed of 100 mm/s, with a simultaneous echocardiogram and phonocardiogram. The patients were studied in either the supine or left lateral decubitus, with the

¹British Heart Foundation Research Fellow.

Received for publication 5 August 1977

transducer in the 3rd or 4th left intercostal space. Simultaneous echoes from the left surface of the interventricular septum and the posterior left ventricular wall were obtained at the level of the tips of the mitral leaflets. Only high quality tracings showing clear continuous echoes throughout the cardiac cycle were used for analysis.

Ejection fraction, mean velocity of circumferential fibre shortening, and normalised left ventricular posterior wall velocity were determined manually by direct measurement of the echocardiograms as previously described (Karliner *et al.*, 1971; Fortuin *et al.*, 1972; Quinones *et al.*, 1974; Feigenbaum and Chang, 1976).

Maximal diastolic endocardial velocity was measured by the method described by Kovick *et al.* (1975) by drawing a tangent to the steepest portion of the diastolic endocardial excursion and measuring the slope in cm/s (Fig. 1).

DIGITISATION

Echocardiograms were then digitised as described by Gibson and Brown (1973). Briefly, echoes from the left surface of the interventricular septum, posterior wall, and anterior mitral leaflet were traced by a cursor. Strings of co-ordinates were generated for approximately 100 points from each of these echoes. This information, together with calibration signals representing 0.5 s, 1 cm, and RR

interval of the beat being studied, were stored on an IBM computer for subsequent calculations.

From the stored digitised data, plots were made of the position of the posterior wall, septum and anterior mitral leaflet, instantaneous mitral leaflet velocity, instantaneous left ventricular dimension, the rate of change of dimension, and the normalised rate of change of dimension or the lengthening rate (Fig. 2). Peak velocities of the mitral leaflet movement during early diastolic opening and mid-diastolic closure were measured directly from the plots, as were the end-diastolic dimensions, end-systolic dimensions, peak lengthening rates, and peak shortening rates. Assessment of the relation of wall movement to mitral valve movement was made by measurement of the intervals between the time of (1) the onset of outward wall movement and that of the mitral leaflet opening movement, and (2) the peak rate of change of dimension and peak opening velocity of the mitral leaflet. The time interval from the aortic component of the second heart sound to the onset of mitral valve movement (isovolumic relaxation period) was also measured.

Results

NORMAL

The values for this group of subjects are shown in Tables 1 and 2.

Fig. 1 Echocardiogram from a patient with Duchenne's muscular dystrophy showing measured maximal diastolic endocardial velocity (10.5 cm/s). IVS, interventricular septum; LVW, left ventricular posterior wall; Endo, endocardium; PGC, phonocardiogram; ECG, electrocardiogram.

Fig. 2 Digitised echocardiogram from a patient with Duchenne's muscular dystrophy. The lowest panel represents septal, mitral valve, and posterior wall echoes. Above are plotted left ventricular dimension, normalised lengthening rate, and anterior mitral valve cusp velocity.

DUCHENNE'S MUSCULAR DYSTROPHY

Systolic indices (Table 3)

Ejection fraction, mean velocity of circumferential fibre shortening, maximal systolic endocardial velocity, and normalised posterior wall velocity were all within the normal range.

Diastolic indices (Table 4)

The peak lengthening rates and peak rates of change of dimension were not significantly different from normal. In addition, the pattern of mitral valve movement followed closely that of wall movement in the normal fashion. The onset of outward wall movement occurred within 2.31 ± 5.9 ms of the onset of anterior movement of mitral valve. Similarly, the peak rate of outward wall movement occurred within 17.7 ± 20.2 ms of the peak rate of mitral valve

opening velocity. Neither of these intervals is significantly different from normal.

In contrast, the maximal diastolic endocardial velocity determined manually was reduced in patients with Duchenne's muscular dystrophy. In 3 patients the value was outside the normal range and the mean value was significantly different from normal ($P < 0.01$). There was no clear relation between abnormality of the maximal diastolic endocardial velocity and the severity of skeletal muscle disorder. It was not possible to predict this echocardiographic abnormality from the presence of abnormal features on the electrocardiogram, analysed in Table 5.

Discussion

Cardiac complications in Duchenne's muscular dystrophy have been reported by different authors, the frequency of complications depending on the age group studied and the criteria used. Myocardial involvement in Duchenne's muscular dystrophy was first noted in 1883 by Ross who described myocardial atrophy in a 12-year-old boy with pseudohypertrophic paralysis. Globus (1923) suggested that histological changes in the heart were in fact cardiac manifestations of systemic dystrophy. Later authors have shown that histological changes in the myocardium were similar to those in the skeletal muscle though less diffuse and were present in all the patients examined at necropsy (Bevans, 1945; Moore, 1954; Schott *et al.*, 1955; Dubowitz, 1960; Watson, 1968; Dubowitz, 1978). There seems little doubt that myocardial involvement is common in Duchenne's muscular dystrophy, though it is not known at what stage during the disease myocardial involvement first occurs, but the electrocardiographic changes are often present in the young, suggesting an early involvement. No correlation has, however, been found between the degree of skeletal muscle disease, severity of cardiac symptoms, and electrocardiographic abnormalities (Cannon, 1962), and this lack of correlation was confirmed in this study.

The earliest indication of dystrophic heart disease has been the appearance of electrocardiographic abnormalities seen in 50 to 90 per cent of cases of Duchenne's muscular dystrophy (Rubin and Buchberg, 1952; Berenbaum and Horowitz, 1956; Manning and Cropp, 1958; Dubowitz, 1960; Gilroy *et al.*, 1963; Wahi, 1963; Perloff *et al.*, 1967; Dubowitz, 1978). The most frequent electrocardiographic abnormalities reported have been tachycardia (Boas and Lowenberg, 1931; Manning and Cropp, 1958; Levin *et al.*, 1959), tall R waves in right precordial leads, with deep Q waves in limb leads and/

Table 1 Systolic function in normal subjects

No.	Age (y)	RR interval	ESD (cm)	SEVM (cm/s)	VPW s ⁻¹	EF (%)	mVcf (circ/s)	Peak Vcf (cm)
1	8	0.5	2.6	5.6	1.22	64	1.4	3.0
2	7	0.6	2.3	8.7	1.04	89	2.0	4.0
3	5	0.7	2.3	5.3	1.16	64	1.3	3.0
4	7	0.5	2.3	6.7	0.95	75	1.4	4.0
5	9	0.7	1.8	8.0	1.41	86	2.1	6.0
6	7	0.8	1.6	6.8	1.28	77	1.5	3.0
7	8	0.6	2.6	7.2	1.11	70	1.4	4.0
8	11	0.5	2.6	6.3	1.08	69	1.3	4.0
9	9	—	—	6.0	0.97	80	1.9	—
10	10	0.8	2.5	5.2	0.91	80	1.4	3.5
11	12	0.8	2.3	5.2	0.81	74	1.4	3.5
12	6	0.6	2.3	—	—	—	—	4.0
13	6	0.6	2.3	6.1	0.81	72	1.4	4.0
14	12	1.1	2.3	6.8	0.81	78	1.3	3.0
15	5	0.8	2.3	6.8	1.09	80	1.7	3.5
16	7	0.5	2.4	7.5	1.06	77	1.7	4.0
17	5	0.6	2.0	5.0	1.02	81	1.7	3.5
Mean ±SD	8.9	0.66 ±0.17	2.3 ±0.3	6.4 ±1.0	1.06 ±0.15	7.5 ±7.1	1.6 ±0.3	3.6 ±0.8

Abbreviations: ESD, end-systolic dimension; SEVM, maximal systolic endocardial velocity; VPW, normalised posterior left ventricular wall velocity; EF, ejection fraction; mVcf, mean velocity of circumferential fibre shortening; peak Vcf, peak velocity of fibre shortening (peak shortening rate).

Table 2 Diastolic function—normal subjects

No.	Age (y)	EDD (cm)	Peak dD/dt	Peak lengthening rate (s ⁻¹) (dD/dt/D)	D _{min} -MVO _{ms}	A ₁ -MVO (ms)	Peak MVO velocity (D-E) cm s ⁻¹	Peak MVC velocity (EF) cm s ⁻¹	Peak MVO-dD/dt/D ms	DEVm cm/s
1	8	3.8	13.2	5.5 (2.4)	0	5	30	20	0	14.0
2	7	4.0	13.7	5.0 (2.7)	20	20	80	20	0	18.3
3	5	3.5	16.2	6.0 (2.7)	0	25	40	30	0	13.5
4	7	3.7	16.2	6.0 (2.7)	0	0	30	20	0	13.4
5	9	3.5	22.0	9.5 (2.3)	10	40	80	30	10	22.0
6	7	3.7	13.2	6.0 (2.2)	10	30	50	20	0	12.1
7	8	3.7	11.2	4.0 (2.8)	0	5	60	30	25	20.0
8	11	4.0	12.6	4.5 (2.8)	0	0	80	20	20	17.1
9	9	—	—	—	—	—	—	—	—	12.7
10	10	4.0	14.0	5.0 (2.8)	0	0	55	20	35	15.4
11	12	4.0	15.0	5.5 (2.8)	0	0	70	30	15	14.5
12	6	4.0	13.7	5.0 (2.7)	0	0	50	20	0	19.7
13	6	4.0	21.0	7.5 (2.8)	5	5	75	25	0	15.2
14	12	4.5	9.3	3.0 (3.1)	0	0	50	15	0	14.2
15	5	4.0	13.2	5.0 (2.6)	0	0	80	25	50	10.2
16	7	3.9	14.0	5.0 (2.8)	0	10	60	35	0	12.6
17	5	3.6	11.5	5.0 (2.3)	0	0	30	15	25	15.9
Mean ±SD	8.9	3.8 ±0.3	14.1 ±3.5	5.3 ±1.6	2.81 ±5.76	9.33 ±13.07	55.6 ±17.8	23.4 ±6.0	12.9 ±16.1	15.3 ±3.1

Abbreviations: EDD, end-diastolic dimension; Peak dD/dt, peak rate of change of dimension; D_{min}-MVO_{ms}, time interval from minimum dimension to onset of mitral valve opening; A₁-MVO, time interval from aortic valve closure to mitral valve opening (isovolumic relation period); MVO, mitral valve opening; MVC, mitral valve closing; DEVm, maximal diastolic endocardial velocity; peak MVO-dD/dt/D, time interval from peak mitral valve opening to peak rate of change of dimension; —, data not available.

or left praecordial leads (Weisenfeld and Messinger, 1952; Manning and Cropp, 1958; Gilroy *et al.*, 1963; Wahi, 1963; Perloff *et al.*, 1966). Rarely, left ventricular hypertrophy may occur (Keith *et al.*, 1967). Conduction defects and non-specific T wave changes are also seen. It has been suggested that the anterior shift of the QRS (tall right praecordial R waves) relates to a loss of posteriorly directed electrical forces caused by selective scarring of the posterobasal portion of the left ventricle. The abnormally deep Q waves appear to reflect lateral extension of this scarring (Perloff *et al.*, 1967).

Histologically, the cardiac lesion consists of fatty and fibrous tissue replacement of the myocardium with selective scarring of the posterolateral wall of the left ventricle and sometimes the posterolateral papillary muscle (Nothacker and Netsky, 1950; Moore, 1954; Schott *et al.*, 1955; Storstein and Austraheim, 1955; Dubowitz, 1960; Perloff *et al.*, 1967; Dubowitz, 1978). The large coronary arteries are normal. Though a non-inflammatory degenerative arteriopathy of small vessels has been described by James (1962), the distribution of scarring is unrelated to the arteriopathy (Perloff *et al.*, 1967). The

Table 3 Systolic function—patients

No.	Age (y)	RR interval	ESD (cm)	SEVM (cm/s)	VPW s ⁻¹	EF (%)	mVcf circ/s	Peak Vcf cm s ⁻¹
1	11	0.60	2.3	7.0	1.22	67	1.3	3.5
2	8	0.42	2.0	7.5	1.19	81	2.0	5.0
3	12	0.52	2.3	6.4	1.08	68	1.4	3.0
4	9	0.60	2.3	3.8	0.84	67	1.2	3.0
5	10	0.40	2.5	5.6	1.02	73	1.7	4.0
6	6	0.45	3.5	6.3	0.88	66	1.4	5.2
7	11	0.65	2.0	4.5	1.10	70	1.5	4.0
8	8	0.60	2.7	7.5	1.03	70	1.4	4.0
9	9	0.50	2.6	3.6	0.82	74	1.4	4.0
10	7	0.55	2.3	3.8	0.95	75	1.6	3.0
11	10	0.60	2.3	6.1	1.01	59	1.3	5.0
12	8	0.50	2.4	7.4	1.32	62	1.3	4.0
13	5	0.40	2.0	4.0	0.90	65	1.3	4.0
Mean ±SD	8.7	0.52 ±0.09	2.4 ±0.4	5.6 ±1.5	1.00 ±0.16	69.7 ±5.2	1.5 ±0.2	4.0 ±0.8

Abbreviations: ESD, end-systolic dimension; SEVM, maximal systolic endocardial velocity; VPW, normalised posterior left ventricular wall velocity; EF, ejection fraction; mVcf, mean velocity of circumferential fibre shortening. Peak Vcf, peak velocity of fibre shortening (peak shortening rate).

Table 4 Diastolic function—patients

No.	Age (y)	EDD (cm)	Peak dD/dt	Peak lengthening rate s ⁻¹ (dD/dt/D)	Dmitr-MVO _{ms}	A ₁ -MVO ms	Peak MVO velocity (D-E) cm s ⁻¹	Peak MVC velocity (EF) cm s ⁻¹	Peak MVO dD/dt/D ms	DEVCM cm/s
1	11	3.2	10.4	4.0 (2.6)	0	5	50	40	0	12.6
2	8	3.5	13.2	6.0 (2.2)	0	0	40	20	10	10.2
3	12	4.1	9.3	3.0 (3.1)	0	0	80	20	0	12.1
4	9	3.7	13.5	4.5 (3.0)	0	5	60	30	20	13.3
5	10	4.0	13.0	4.5 (2.9)	0	5	80	35	50	9.8
6	6	5.2	16.0	4.0 (4.0)	0	0	50	40	50	6.0
7	11	3.0	18.0	9.0 (2.0)	0	0	30	20	40	10.0
8	8	4.2	18.6	6.0 (3.1)	0	0	40	30	15	16.4
9	9	4.0	11.2	4.0 (2.8)	0	0	80	20	40	7.6
10	7	3.7	20.0	7.0	0	10	77	20	0	11.9
11	10	3.6	13.5	5.0 (2.7)	10	10	65	20	5	7.2
12	8	3.7	16.2	6.0 (2.7)	0	0	30	20	0	17.0
13	5	3.3	16.1	7.0 (2.3)	0	10	100	15	0	10.2
Mean ±SD	8.7	3.7 ±0.5	15.0 ±2.8	5.6 ±1.5	2.31 ±4.37	4.09 ±4.37	59.6 ±20.3	25.8 ±8.7	17.7 ±20.2	11.1 ±3.2

Abbreviations (as in Table 2).

Table 5 Analysis of electrocardiogram of patients

No.	Age (y)	HR (beats/min)	Rhythm	QRS frontal axis	P wave	PR interval (s)	Deep Q wave	Tall right praecordial R waves	ST segment	T wave	LVH
1	11	120	SR	+45	N	0.12	I, VL, V4-6	—	N	N	+
2	8	100	Junctional	+30	Inverted in II, III, VF	0.08	V5 and V6	—	N	N	+
3	12	100	SR	+60	N	0.12	I, VL, V4-6	+	N	N	—
4	9	95	SR	+45	N	0.14	—	—	N	N	—
5	10	90	SR	+80	N	0.12	—	+	N	N	—
6	6	100	SR	+45	N	0.12	—	+	N	Flat in V4 and V5	—
7	11	110	SR	+30	N	0.12	—	+	N	N	—
8	8	90	SR	+120	N	0.10	—	+	N	N	—
9	9	100	SR	+120	N	0.14	II, III, VF, and V4-6	—	N	N	—
10	7	105	SR	+90	N	0.12	—	+	N	N	—
11	10	130	SR	+60	N	0.14	I, VL, V4-6	—	N	N	—
12	8	130	SR	+60	N	0.08	V4-6	+	N	N	—
13	5	120	SR	+80	N	0.12	—	—	—	—	—

Abbreviations: HR, heart rate; LVH, left ventricular hypertrophy; SR, sinus rhythm; +, present; —, absent; N, normal.

unusual focal location of myocardial fibrosis can not be explained but correlates well with the common electrocardiographic abnormalities.

This association led Kovick *et al.* (1975) to examine posterior left ventricular wall motion using echocardiography in patients with all types of progressive muscular dystrophy including 3 patients (ages 15, 16, and 22) with Duchenne's muscular dystrophy. They found a reduced maximal diastolic endocardial velocity and suggested that this reflected impaired myocardial relaxation caused by dystrophic heart disease. Our results have confirmed their findings in showing that there was a significant difference in the mean of the Duchenne muscular dystrophy group as compared with the normal group. However, the maximal diastolic endocardial velocity was found to be normal in some patients who had electrocardiographic evidence of myocardial involvement. Therefore, though this index may reflect dystrophic heart disease, it is not a sensitive index of myocardial involvement in individual patients. Our observations, however, do not necessarily conflict with those of Kovick *et al.* (1975), who suggested that a reduced maximal diastolic endocardial velocity might be a sensitive index of myocardial involvement in muscular dystrophy but studied only 3 patients with Duchenne's muscular dystrophy, all of whom had electrocardiographic abnormalities. We are not clear as to what causes the reduced maximal diastolic endocardial velocity since the ventricle is filling at a normal rate as witnessed by normal peak lengthening rates. The measurement of movement of a surface (interface) such as endocardium seen echocardiographically is influenced by overall movement of the heart within the thorax. A reduction of motion of the whole heart from whatever cause tends to reduce the velocity of the endocardium measured directly from the echocardiogram. While normal peak rate of change of dimension would be preserved since there is progressive involvement of respiratory muscles in Duchenne's muscular dystrophy, this results in reduced overall motion of the heart within the thorax. Sandberg *et al.* (1952) noted reduced excursion of cardiac motion on fluoroscopic examination of patients with Duchenne's muscular dystrophy.

Diastole in the normal heart is a well co-ordinated and precisely timed process. Echocardiographic studies have shown that the movement of the left ventricular wall is closely related to the movement of the mitral valve cusps with both events having an identical pattern of movement. Disturbance of this close relation seems to be a sensitive index of myocardial disorder, and abnormalities have been shown both in ischaemic heart disease and hyper-

trophic obstructive cardiomyopathy (Altieri *et al.*, 1973; Ruttlely *et al.*, 1974; Wilson *et al.*, 1975; Gibson *et al.*, 1976; Upton *et al.*, 1976).

The maintenance of this close time relation in the patients studied here suggests that myocardial function is well preserved in Duchenne's muscular dystrophy despite electrocardiographic changes. It is possible that the involvement of the cardiac muscle is not so extensive and severe as to cause cardiac dysfunction but sufficiently localised to modify the electrocardiographic pattern.

In conclusion, an echocardiographic study in children with Duchenne's muscular dystrophy has shown that myocardial function appears to be well preserved despite electrocardiographic abnormalities. Previously noted reduced maximal diastolic endocardial velocity is confirmed in a more homogeneous and a larger group of patients but may reflect a reduced overall movement of the heart within the thorax rather than a functional cardiac abnormality.

We thank Dr Celia M. Oakley and Professor J. F. Goodwin for their advice, and Dr David Brown (Brompton Hospital) for his assistance in computer analysis.

References

- Altieri, P. I., Wilt, S. M., and Leighton, R. F. (1973). Left ventricular wall motion during the isovolumic relaxation period. *Circulation*, **48**, 499-505.
- Berenbaum, A. A., and Horowitz, W. (1956). Heart involvement in progressive muscular dystrophy; report of a case with sudden death. *American Heart Journal*, **51**, 622-627.
- Bevans, M. (1945). Changes in the musculature of the gastrointestinal tract and in the myocardium in progressive muscular dystrophy. *Archives of Pathology*, **40**, 225-238.
- Boas, E. P., and Lowenberg, H. (1931). The heart rate in progressive muscular dystrophy: studies with the cardiometer. *Archives of Internal Medicine*, **47**, 376-383.
- Cannon, P. J. (1962). The heart and lungs in myotonic muscular dystrophy. *American Journal of Medicine*, **32**, 765-775.
- Dubowitz, V. (1960). Progressive muscular dystrophy in childhood. M.D. Thesis, University of Cape Town.
- Dubowitz, V. (1978). *Muscle Disorders in Children*. Saunders, London and Philadelphia.
- Feigenbaum, H., and Chang, S. (1976). *Echocardiography*, pp. 297-340. Lea and Febiger, Philadelphia.
- Fortuin N. J., Hood, W. P., Jr., and Graige, E. (1972). Evaluation of left ventricular function by echocardiography. *Circulation*, **46**, 26-35.
- Gibson, D. G., and Brown, D. J. (1973). Measurement of instantaneous left ventricular dimension and filling rate in man using echocardiography. *British Heart Journal*, **35**, 1141-1149.
- Gibson, D. G., Prewitt, T. A., and Brown, D. J. (1976). Analysis of left ventricular wall movement during isovolumic relaxation and its relation to coronary artery disease. *British Heart Journal*, **38**, 1010-1019.
- Gilroy, J., Cahalan, J. L., Berman, R., and Newman, M. (1963). Cardiac and pulmonary complications in

- Duchenne's progressive muscular dystrophy. *Circulation*, **27**, 484-493.
- Globus, J. H. (1923). The pathologic findings in the heart muscle in progressive muscular dystrophy. *Archives of Neurology and Psychiatry (Chicago)*, **9**, 59-72.
- James, T. N. (1962). Observations on the cardiovascular involvement, including the cardiac conducting system, in progressive muscular dystrophy. *American Heart Journal*, **63**, 48-56.
- Karliner, J. S., Gault, J. H., Eckberg, D., Mullins, C. B., and Ross, J., Jr. (1971). Mean velocity of fiber shortening; a simplified measure of left ventricular myocardial contractility. *Circulation*, **44**, 323-333.
- Keith, J. D., Rowe, R. D., and Vlad, P. (1967). *Heart Disease in Infancy and Childhood*, 2nd ed., p. 1138. MacMillan, New York.
- Kovick, R. B., Fogelman, A. M., Abbasi, A. S., Peter, J. P., and Pearce, M. L. (1975). Echocardiographic evaluation of posterior left ventricular wall motion in muscular dystrophy. *Circulation*, **52**, 447-454.
- Levin, S., Baens, G. S., and Weinberg, T. (1959). Heart in pseudohypertrophic muscular dystrophy. *Journal of Pediatrics*, **55**, 460-472.
- Manning, G. W., and Cropp, G. J. (1958). The electrocardiogram in progressive muscular dystrophy. *British Heart Journal*, **20**, 416-420.
- Moore, W. F., Jr. (1954). Cardiac involvement in progressive muscular dystrophy. *Journal of Pediatrics*, **44**, 683-687.
- Nothacker, W. A., and Metsky, M. G. (1950). Myocardial lesion in progressive muscular dystrophy. *Archives of Pathology*, **50**, 578-600.
- Perloff, J. K., De Leon, A. C., Jr., and O'Doherty, D. (1966). The cardiomyopathy of progressive muscular dystrophy. *Circulation*, **33**, 625-648.
- Perloff, J. K., Roberts, W. C., De Leon, A. C., Jr., and O'Doherty, D. (1967). The distinctive electrocardiogram of Duchenne's progressive muscular dystrophy. *American Journal of Medicine*, **42**, 179-188.
- Quinones, M. A., Gaasch, W. H., and Alexander, J. K. (1974). Echocardiographic assessment of left ventricular function— with special reference to normalised velocities. *Circulation*, **50**, 42-51.
- Ross, J. (1883). On a case of pseudohypertrophic paralysis. *British Medical Journal*, **1**, 200-202.
- Rubin, I. L., and Buchberg, A. S. (1952). The heart in progressive muscular dystrophy. *American Heart Journal*, **43**, 161-169.
- Ruttley, M. S., Adams, D. F., Cohn, P. F., and Abrams, H. (1974). Shape and volume changes during 'isovolumic relaxation' in normal and asynergic ventricles. *Circulation*, **50**, 306-316.
- Sandberg, A. A., Hecht, H. H., and Tyler, F. H. (1952). The heart in muscular dystrophy (abstract). *American Journal of Medicine*, **13**, 498-499.
- Schott, J., Jacobi, M., and Wald, M. A. (1955). Electrocardiographic patterns in the differential diagnosis of progressive muscular dystrophy. *American Journal of the Medical Sciences*, **229**, 517-524.
- Storstein, O., and Austraheim, K. (1955). Progressive muscular dystrophy of the heart. *Acta Medica Scandinavica*, **150**, 431-436.
- Upton, M. T., Gibson, D. G., and Brown, D. J. (1976). Echocardiographic assessment of abnormal left ventricular relaxation in man. *British Heart Journal*, **38**, 1001-1009.
- Wahi, P. L. (1963). Cardiac change in myopathy. *American Heart Journal*, **66**, 748-754.
- Watson, H. (1968). *Paediatric Cardiology*, p. 770. Lloyd-Luke, London.
- Weisenfeld, S., and Messinger, W. J. (1952). Cardiac involvement in progressive muscular dystrophy. *American Heart Journal*, **43**, 170-187.
- Wilson, C. S., Krueger, S., Forker, A. D., and Weaver, W. F. (1975). Correlation between segmental early relaxation of the ventricular wall and coronary occlusive disease. *American Heart Journal*, **89**, 474-479.

Requests for reprints to Dr K. A. Hallidie-Smith, Royal Postgraduate Medical School, Hammersmith Hospital, Du Cane Road, London W12 OHS.