Cardiorespiratory changes in progressive muscular dystrophy

P. L. Wahi, K. C. Bhargava, and S. Mohindra

From the Department of Medicine, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Incipient cardiac failure was detected in 56 per cent of cases of pseudohypertrophic muscular dystrophy, 50 per cent of limb girdle type of myopathies, and 2 of the 3 cases with 'unclassified' variety.

'Myopathic pattern' in the electrocardiogram was seen in 82 per cent of the cases of pseudohypertrophic muscular dystrophy who showed abnormal haemodynamics. This suggests that this pattern is due to myocardial damage. Thus the electrocardiogram can be used as a simple and safe procedure for the detection of dystrophic heart disease.

Though 'myopathic pattern' is seen only in pseudohypertrophic muscular dystrophy the haemodynamic evidence of dystrophic heart disease is seen in other myopathies as well.

Pulmonary ventilatory function tests revealed restrictive changes in 57 per cent of cases of myopathy when muscles of the chest wall were involved. This abnormality in pulmonary function tests had no apparent haemodynamic effect.

Cardiopulmonary complications are well known in muscular dystrophy (Globus, 1923; Nothacker and Netsky, 1950; Hurwitz, 1936). They occur in the form of cardiac arrhythmias, congestive cardiac failure, electrocardiographic changes, pulmonary infections, and pulmonary insufficiency (Rubin and Buchberg, 1952; Weisenfeld and Messinger, 1952; Manning and Cropp, 1958; Gilroy *et al.*, 1963; Arce-Gomez *et al.*, 1964).

A specific 'myopathic pattern' in the electrocardiogram in cases with pseudohypertrophic muscular dystrophy has been described by us. This pattern consists of sinus tachycardia, short PR interval, R/S in V₁ more than 1.5, high amplitude R wave and deep Q wave in V₄₋₆ and other left precordial leads, and prolonged QT_c (Fig. 1). It was suggested that these electrocardiographic changes were probably due to myocardial damage (Wahi, 1961, 1963).

Detailed pulmonary and haemodynamic studies in progressive muscular dystrophy are few (Gailani, Danowaki, and Fisher, 1958; Storstein, 1964; Perloff, de Leon, and O'Doherty 1966; Perloff *et al.*, 1967; Gilroy *et al.*, 1963; Wahi *et al.*, 1968; Arce-Gomez *et al.*, 1964). The incidence of pulmonary infections, mostly pneumonias, is high in advanced cases of myopathy. Ventilatory function studies in these cases have shown

restrictive changes with limitation of all lung volumes. In our total experience of 180 cases of myopathy we have encountered bronchiectasis in two cases, bronchopneumonia in one, and congestive cardiac failure in two cases.

Pulmonary insufficiency and cardiac failure do not manifest early owing to restricted physical activity because of disability. Pulmonary function tests and cardiac catheterization can be useful means of detecting early changes and latent heart failure.

Material and methods

The present study is of 41 cases of progressive muscular dystrophy. Out of a large series of 180 cases of myopathy 32 were studied haemodynamically in the Nehru Hospital of the Postgraduate Institute of Medical Education and Research, Chandigarh, and 9 were studied for us by Professor S. B. Roy at All India Institute of Medical Sciences, New Delhi, before our own laboratories were set up. There were 30 cases with pseudohypertrophic muscular dystrophy (sex-linked, Duchenne type), 8 cases of limb girdle myopathy, and another 3 cases were atypical and were grouped as 'unclassified'. Muscle biopsy was done in all cases where the clinical picture was even slightly atypical (14 cases). After detailed clinical examination a radiograph of the chest for heart size and lung fields was done at a standard distance. A 12-lead electrocardiogram was taken in all these cases.

FIG. 1 Showing typical myopathic pattern in electrocardiogram – R/S in V_1 , 1.5, and deep Q waves in lead I, aVL, and V_4 –6.

Ventilatory pulmonary functions were studied in 29 cases. These included tidal volume, minute ventilation, vital capacity, maximum breathing capacity, and forced expiratory volume. In 8 cases functional residual capacity and total lung capacity were also done. All the ventilatory pulmonary functions were done in basal condition, and a repeat study was done where co-operation was inadequate. The pulmonary functions were done by Goddard Pulmotest except functional residual capacity, which was done by open circuit nitrogen wash-out method.

Standard right heart catheterization was done in all these cases. Pressures were recorded at rest, and in suitable cases records were taken after exercise or right hypochondriac compression, or both, to bring out findings of incipient right heart failure. Cardiac output was calculated by the Fick principle.

Findings

The clinical examination found associated atrial septal defect in one patient and another had mild hypertension. None of these patients

showed frank congestive cardiac failure or any physical signs in the chest. Radiography showed cardiac enlargement in 4 cases. There was no pulmonary pathology. Twenty out of 30 patients (66%) with pseudohypertrophic muscular dystrophy showed a 'myopathic pattern' in the electrocardiogram (Wahi, 1963).

Haemodynamic studies Twenty-seven patients had normal values for resting pressures. Twelve had abnormal right ventricular end-diastolic pressure and in another two cases the pressures were on higher limits of normal (6 mm.) (Fig. 2). Leg-raising exercise for three minutes could be given in 6 patients, and only one of these showed distinct elevation of right ventricular end-diastolic pressure (Fig. 3). Right hypochondriac compression was applied in 32 patients. Eleven patients with normal or borderline resting pressure showed elevation of right ventricular end-diastolic pressure after right hypochondriac compression (Fig. 4). Generally the patients having abnormal right ventricular end-diastolic pressure at rest also had abnormal mean right atrial pressure. Right atrial mean pressure showed an increase corresponding to changes in right ventricular end-diastolic pressure after right hypochondriac compression.

FIG. 2 Showing abnormal right ventricular end-diastolic pressure at rest.

FIG. 3 Showing normal right ventricular end-diastolic pressure at rest and its abnormal increase after 2 min exercise.

FIG. 4 Showing normal right ventricular pressures at rest and abnormal increase of right ventricular end-diastolic pressure after right hypochondriac compression and return to normal after release of pressure.

sion except in two cases (Fig. 5). In total, 23 out of 41 patients (56%) showed abnormal haemodynamic findings suggestive of cardiac failure. This was noticed in 17 out of 30 cases of pseudohypertrophic muscular dystrophy (56%), 4 out of 8 cases of limb girdle myopathy (50%), while 2 out of 3 cases of unclassified variety had abnormal haemodynamics.

Pulmonary artery pressure was normal in all, and there was no systolic gradient across the pulmonary valve. Pulmonary capillary pressure was found to be normal in all except one case with mild hypertension.

Cardiac output was measured in 36 cases. It varied from 3–6.6 l./min. Cardiac index was normal in 28 cases (3–5 l./min/m²), 2 cases showed a rise (5.5, 7.7 l./min/m²), and 5 showed decreased values (less than 2.5 l./min/m²).

Considerable irritability of the myocardium was noticed in the form of arrhythmias during catheterization as compared to the other patients studied in the same laboratory during the same period. One patient with a high right

ventricular end-diastolic and mean right atrial pressure developed ventricular tachycardia during catheterization but was successfully revived.

Pulmonary function studies The details of the ventilatory pulmonary function tests are described in different groups separately (Tables 1, 2, 3). Vital capacity and maximum breathing capacity were reduced in two-thirds

TABLE 1 *Pseudohypertrophic muscular dystrophy*

Case No.	Vital capacity (l.)	% of predicted vital capacity	Maximum breathing capacity (l.)	% of predicted maximum breathing capacity	FEV ₁
2	0.840	40	30.6	50.1	82
3	0.930	40.8	24.3	34.6	71
4	1.170	46.8	38.4	52.6	89
5*	1.050	40.3	—	—	77
6	0.900	47.36	23.4	42.5	90
7	3.120	78	90.82	73.2	92
8	1.980	79.2	55.2	81.1	92
9	3.18	88.2	93.06	90.3	80.2
12	3.60	94.7	118.8	104.8	83
13	0.720	30	20.16	28	95
15	1.14	57	34.56	40.6	84.7
16*	0.810	41.5	28.56	51.8	88.9
18*	1.08	46.9	30.24	43.2	—
19*	1.20	40	37.44	40.6	95
22*	0.870	44.6	—	—	90
28	1.080	42	30.60	39.0	—
29	4.050	100	124.800	100	89

* Co-operation was not adequate.

TABLE 2 *Limb girdle type*

Case No.	Vital capacity (l.)	% of predicted vital capacity	Maximum breathing capacity (l.)	% of predicted maximum breathing capacity	FEV ₁
1	3.55	87.5	88.5	84.9	50
10	3.75	89.2	96.5	84.5	76
11	3.27	102.5	122.78	113	70
14	2.25	54.8	60.48	62.6	88
17	2.25	56.2	67.20	62.8	82
20	2.25	62.5	98.84	90.2	93
23	3.81	74.6	96.36	69	94
24	3.03	70	101.76	92.4	94

TABLE 3 *Unclassified variety*

Case No.	Vital capacity (l.)	% of predicted vital capacity	Maximum breathing capacity (l.)	% of predicted maximum breathing capacity	FEV ₁
25	3.66	88	92.4	88.3	75
26	2.61	103	75.6	102	80
27	3.00	100	84.4	88	93

FIG. 5 Showing normal resting mean right atrial pressure, its increase after right hypochondriac compression, and return to normal after release of pressure.

of the cases of pseudohypertrophic muscular dystrophy. Similar changes were seen in 44.4 per cent of limb girdle type of myopathy, but the cases with 'unclassified' variety of myopathy had normal values. Timed vital capacity (FEV₁) did not suggest any obstructive airway disease. Functional residual capacity and total lung capacity were done in 8 cases and were found to be normal in 4 cases which showed other function to be normal. Four cases had reduced values, and they also showed restrictive airway disease as judged from vital capacity and maximum breathing capacity.

Discussion

Fifty-six per cent of cases of myopathy studied by us showed raised right ventricular end-diastolic or mean right atrial pressure, or both, at rest or when challenged with exercise or right hypochondriac compression, though none of them had shown clinical evidence of right heart failure. A rise of right ventricular end-diastolic pressure or mean right atrial pressure, or both, after abdominal compression may reflect reduced distensibility of the right ventricle and its inability to handle augmented volume overloading.

Out of 17 cases of Duchenne type of muscular dystrophy who showed abnormal haemodynamics, 14 (82%) showed typical 'myopathic pattern' in the electrocardiogram. This supports our contention that this pattern is an evidence of dystrophic heart disease and not a conduction defect. Similar studies by Perloff *et al.* (1966) also support this hypothesis.

However, 'myopathic pattern' was seen in 20 cases of pseudohypertrophic muscular dystrophy, while haemodynamic abnormalities were seen in 17 only. Besides, some patients with abnormal haemodynamics did not show the characteristic electrocardiographic changes. It is possible that the electrocardiographic pattern is suggestive of predominantly localized dystrophic changes in the myocardium in the region of the posterolateral wall of the left ventricle which may occur earlier than the disturbed haemodynamics. The cases with more diffuse damage may not show characteristic electrocardiographic changes but depict abnormal haemodynamics. Such a view is supported by the observation of Perloff *et al.* (1966, 1967). Our study suggests that, though 'myopathic pattern' in the electrocardiogram was limited to the cases of pseudohypertrophic muscular dystrophy, abnormal haemodynamics and thus the 'dystrophic heart disease' is equally

common in limb girdle and unclassified varieties.

The ventilatory pulmonary function tests showed that 57 per cent of cases had changes suggestive of restrictive airway disease, which were present in Duchenne and limb girdle type of myopathies. These changes were not seen in the unclassified variety because of the predominantly distal muscular involvement. The ventilatory function abnormality is due to inadequate expansion of lungs because of weakness of the respiratory muscles. In spite of these restrictive changes there were no symptoms of respiratory insufficiency, most probably because of limited physical activity due to generalized muscular weakness. None of the cases showed any evidence of obstructive airway disease. The changes in the pulmonary functions had no apparent haemodynamic effect, as pulmonary artery pressure and peripheral oxygen saturation were normal in all these cases.

We are grateful to Professor J. N. Berry and his team for doing some of the haemodynamic studies.

References

- Arce-Gomez, E., Palma-Garcia, S., Yanez, J. L., Lopez-Portillo, M., Lombardo, L., Brandt, H., and Marquez, M. H. (1964). Study of cardio-respiratory function in patients with progressive muscular dystrophy. *Angiology*, **15**, 407.
- Gailani, S., Danowaki, T. S., and Fisher, D. S. (1958). Muscular dystrophy. Catheterization studies indicating latent congestive heart failure. *Circulation*, **17**, 583.
- Gilroy, J., Cahalan, J. L., Berman, R., and Newman, M. (1963). Cardiac and pulmonary complications in Duchenne's progressive muscular dystrophy. *Circulation*, **27**, 484.
- Globus, J. H. (1923). The pathologic findings in the heart muscle in progressive muscular dystrophy. *Archives of Neurology and Psychiatry*, **9**, 59.
- Hurwitz, S. (1936). Primary myopathies, report of 36 cases and review of the literature. *Archives of Neurology and Psychiatry*, **36**, 1294.
- Manning, G. W., and Cropp, G. J. (1958). The electrocardiogram in progressive muscular dystrophy. *British Heart Journal*, **20**, 416.
- Nothacker, W. G., and Netsky, M. G. (1950). Myocardial lesions in progressive muscular dystrophy. *Archives of Pathology*, **50**, 578.
- Perloff, J. K., de Leon, A. C., and O'Doherty, D. (1966). The cardiomyopathy of progressive muscular dystrophy. *Circulation*, **33**, 625.
- Perloff, J. K., Roberts, W. C., de Leon, A. C., and O'Doherty, D. (1967). The distinctive ECG of Duchenne's progressive muscular dystrophy (an ECG-pathologic correlative study). *American Journal of Medicine*, **42**, 179.
- Rubin, I. L., and Buchberg, A. S. (1952). The heart in progressive muscular dystrophy. *American Heart Journal*, **43**, 161.
- Storstein, O. (1964). The heart in progressive muscular dystrophy. *Experimental Medicine and Surgery*, **22**, 13.

- Wahi, P. L. (1961). Electrocardiographic changes in progressive muscular dystrophy. *Journal of the Association of Physicians of India*, 9, 662.
- Wahi, P. L. (1963). Cardiac changes in myopathy. *American Heart Journal*, 66, 748.
- Wahi, P. L., Manchanda, S. S., Thind, M. S., and Akhtar, M. (1968). Cardiopathy in muscular dystrophy. *Diseases of the Chest*, 53, 79.
- Weisenfeld, S., and Messinger, W. J. (1952). Cardiac involvement in progressive muscular dystrophy. *American Heart Journal*, 43, 170.