Electrocardiographic Studies in the Patients of Duchenne Muscular Dystrophy

Pages with reference to book, From 117 To 121

Naeema Ansari, P.Q.R. Siddiqui (Department of Physiology, University of Karachi.)
S.R. Kirmani (Department of Physical Medicine, Jinnah Postgraduate Medical Centre, Karachi.)

Abstract

Electrocardiographic (ECG) data from twenty Duchenne dystrophic patients aged 6 18 years were examined and compared with normal standards to explore the characteristic ECG abnormalities in these patients. Pathological electrocardiographic patterns were seen in 13 cases whereas 7 electrocardiograms were found normal. Amongst these 13 cases, the ECG of seven patients showed right ventricular hypertrophy (RVH) with strain. The ECG changes of asymmetric septal hypertrophy (ASH) were also observed in these patients. Four patients showed a pattern indicating combined ventricular hypertrophy (CVH) and one had typical low voltage changes (JPMA 34: 117, 1984).

Introduction

Several electrocardiographic (ECG) changes characteristic ef progressive muscular dystrophy were reported by many investigators (Zatuchni et al., 1951; Weisenfield and Messinger, 1952; Wahi, 1963; Emery, 1972; Hunter, 1980; Ishikawa et at., 1982; Sanyal and Johnson, 1982). Among these the tall right precordial R waves, increased ratios of R/S waves and deep Q waves in the lateral and leftward precordial leads are said to be characteristic of muscular dystrophy, particularly of the Duchenne type. Perloff et at. (1966) and Emery (1972) have claimed that this ECG pattern is of diagnostic value in distinguishing the juvenile form of muscular dystrophy. DeLeon et al. (1967) have also shown that the pathological basis for this distinctive ECG pattern in two autopsy cases was the interstitial and replacement fibrosis of the basal part of the left ventricular free wall. The peculiar distribution of the myocardial fibrosis was also reported in muscular dystrophy by Frankle and Rosser (1976). These authors suggested that these pathological findings reflect a generalized metabolic abnormality at cellular level, which becomes obvious initially in the parts of myocardium furthest away from the left ventricular cavity and produces firstly, a loss of epimyocardial forces and subsequently the fibrosis that is so characteristic at autopsy in severely affected hearts. This classical pathological picture, found most commonly in Duchenne dystrophy, differs histologically from the diffuse fibrosis seen in ischaemic heart disease (Hunter, 1980).

Several groups of workers have more recently used the echocardiogram to look at septal and posterior left ventricular wall thickness and movement in patients with muscul dystrophy. Kovick et at. (1975) studied the maximal systolic endocardial velocity (SEVM) and maximal diastolic endocardial velocity (DEYM) in the posterior left ventricular wall of the patients of Duchenne progressive muscular dystrophy, and compared with those in age matched controls, patients with myotonia congenita and patients deconditioned by bed rest. The SE\TM and DEVM of Duchenne patients was found to be significantly less than in other groups. However, the authors suggested that the DEVM from the posterior left ventricular wall is a more accurate detector of cardiac involvement in muscular dystrophy than the SEVM and indeed it is well accepted that both myocardial contraction and relaxation may be significantly abnormal in cardiomyopathic state.

Similar to the Duchenne ECG patterns the carriers of this X-linked recessive disorder also showed abnormal ECG changes. Lukasik (1975) described these changes in 6.6% of the mothers of affected children. Lane et aL, (1980) also observed the changes in density functions for R/s in V1 and R/s in V2

leads in the carriers of Duchenne muscular dystrophy. According to Hunter (1980) the measurement of R/s ratios in the right precordial chest lead seems to be of value in differentiating carriers from control subjects. In the present study we have undertaken an evaluation of scalar electrocardiograms in the patients of Duchenne muscular dystrophy.

Material and Methods

Twenty Duchenne patients (6-18 years age) with classical type of the muscular dystrophy were included in this investigation. These patients were attending the out patient clinic in the Department of Physical Medicine, Jinnah Post-graduate Medical Centre, Karachi and were selected strictly on the basis of the diagnostic criteria for Duchenne muscular dystrophy (Walton and Gardner-Medwin, 1974). The degree of mobilization varied in these patients. Sixteen patients had no restriction of physical activity, two were markedly restricted whereas two patients were moderately restricted. Chest deformities were observed in only three cases.

Standard 12 lead electrocardiograms were obtained for each patient on a Siemen's cardirex instmment at a paper speed of 25mm/sec. and a voltage standardization of 10mm/mv.

Precise electrocardiographit measurements were obtained by using ECG scale and the average measurements of three successive cycles were used to determine the heart rate, rhythm, mean QRS axis and the P-R and Q-T intervals. The electrocardiograms were evaluated by the combination of the methods of Nadas (1958) and Zieglier (1951).

Results

The electrocardiographic findings of 20 patients are presented in Table I and Table II.

Table – 1 Clinical Electrocardiographic Data in Duchenne Muscular Dystrophy.

Patients	Heart Rate (Beats/min)	Axis (Degrees)	P-R Interval (Sec.)	Q-T Interval (Sec.)	QRS Duration (Sec.)
D_3 (10)	80	+ 90	0.17	0.40	0.06
D_4(12)	90	+ 60	0.16	0.32	0.06
D_5 (10)	90	+ 60	0.14	0.38	0.08
D_6 (18)	94	+ 110	0.17	0.28	0.08
D_7 (6)	100	+ 60	0.12	0.26	0.06
D_8(7)	90	+ 60	0.16	0.30	0.04
D_9(14)	103	+ 60	0.16	0.24	0.06
D_{10}(8)	100	+ 60	0.14	0.36	0.06
D_{11}(8)	116	+ 60	0.16	0.36	0.06
D_{16}(10)	102	+ 60	0.10	0.34	0.06
D_{17}(11)	112	+ 30	0.12	0.34	0.06
D_{20}(6)	140	+ 60	0.10	0.30	0.04
D_{22}(10)	120	+ 60	0.08	0.24	0.04
D_{26}(7)	120	+ 60	0.08	0.40	0.06
D_{28}(12)	100	+ 60	0.08	0.44	0.04
D_{32}(18)	100	+ 60	0.12	0.32	0.04
D_{33}(8)	120	+ 60	0.12	0.24	0.04
D_{35}(35)	80	+ 60	0.08	0.32	0.04
D_{36}(11)	120	+ 60	0.12	0.28	0.04
D_{37}(11)	100	+ 60	0.12	0.32	0.04

Figures in parenthesis represent age of the patients.

Table – II

Clinical Electrocardiographic Data in Duchenne Muscular Dystrophy.

Patients	R–VI (m.m.)	S-VI (m.m.)	R/s–V_1	R–V_2 (m.m.)	S–V_2 (m.m.)	R/s–V_2	Q waves (m.m.)	Remarks
D3 (10)	16	12	1.33	23.00	17.00	1.35	Deep:III, avf	RVH with "strain"
D4 (12)	18	15	1.20	20.00	14.00	1.43	Deep: V5, V6	RVH and ASH
D5 (10)	16	05	3.20	15.00	12.00	1.25	Deep: aVL.	RVH with strain
D6(18)	02	05	0.40	12.00	20.00	0.06	Normal	Terminal Delay
D7 (6)	07	08	0.87	06.50	24.00	0.27	Normal	Normal ECG
D8 (7)	15	03	5.00	21.00	14.00	1.50	Deep: 1, aVL,v4,v5, & v6	CVH and ASH
D9(14)	04	11	0.36	09.00	15.50	0.58	Deep:V4, V5	Normal ECG
D10(8)	06	06	1.00	14.00	08.00	1.75	Deep:V3, V4	RVH with strain
D11 (8)	11	09	1.22	23.00	11.00	2.00	Deep:V3, V4, V5 & V6	CVH with strain & ASH
D16 (10)	12	16	0.75	20.00	08.00	2.50	Deep:V4, V5 & V6	Normal ECG
D17(11)	06	15	0.40	10.50	13.00	0.80	Normal	Normal ECG
D20 (6)	02	04	0.50	05.00	04.00	1.25	Not readable	Low voltage
D22 (10)	04	11	0.36	10.00	13.00	0.77	Normal	Normal ECG
D26 (7)	09	06	1.50	22.00	14.00	1.57	Deep : III, aVL	RVH with "strain"
D28 (12)	07	07	1.00	11.00	04.00	2.75	Deep: V5, V6	RVH
D32 (18)	09	05	1.80	15.00	04.00	3.75	Deep:I, aVL, V5, V6	RVH with "strain" & ASH
D33 (8)	09	14	0.64	22.00	20.00	1.10	Normal	Normal
D35 (10)	08	17	0.47	15.00	20.00	0.75	Deep:I,aVL,V4,V5,V6	CVH and ASH
D36 (11)	07	08	0.87	23.00	21.00	1.09	Deep: V4,V5	Normal ECG
D37(11)	29	16	1.81	24.00	21.00	1.14	Deep:II, III, aVR, aVL, aVF, V4, V5, V6.	CVH & ASH

Figures in Parenthesis represent age of patients.

RVH : Right ventricular hypertrophy

CVH : Combined ventricular hypertrophy

ASH : Asymmetric septal hypertrophy

Pathological electrocardiographic patterns were seen in 13 cases whereas seven electrocardiograms were normal. Significantly tall R waves and an abnormally greater R/s ratios were found in right precordial leads in most of the patients. Whereas in five patients R/s ratio in lead V1 was found less than normal for their corresponding ages. These changes were usually accompanied by deep Q waves

not exceeding 3mm in width.
This was commonly observed in leads V5 and V6 and only rarely in the limb leads.
Changes in the T wave and electric axis deviation were rare. Right. axis deviation was found only in two cases (D3 and D6). The Q-T intervals when compared with normal standards, according to the heart rate and cycle length, was found prolonged in six patients. Characteristics of strain pattern were observed in the ECG's of six patients (D3, D5, D10, D11,D26andD32).
Twenty patients with abnormal electrocardiograms were divided into three types according to the different electrocardiographic patterns (Fig. 1).

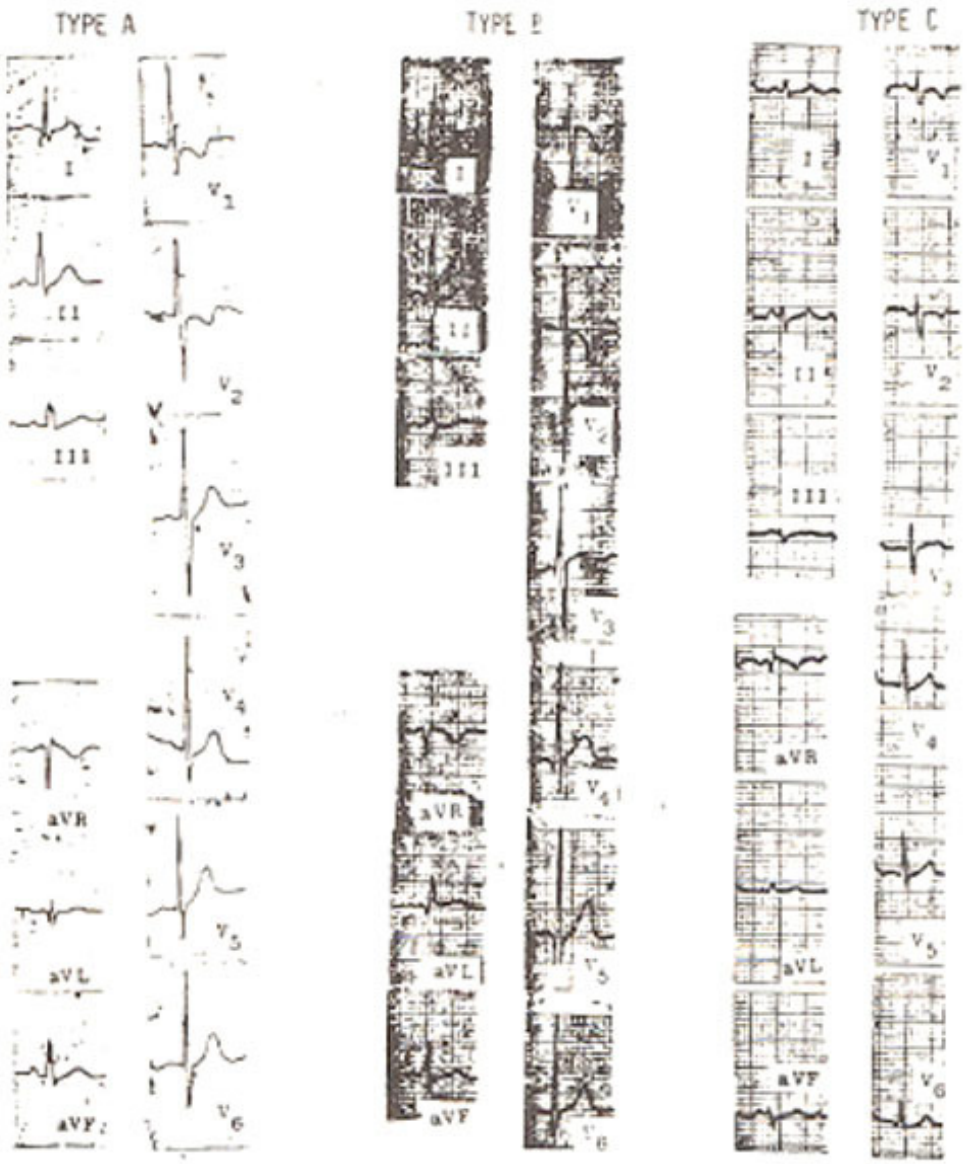

Figure 1. Electrocardiographic Abnormalities in Patients of Duchenne Muscular Dystrophy.

Type A — Right Ventricular Hypertrophy (RVH) with strain and ash in a 10 years old child.

Type B — Combined Ventricular Hypertrophy (CVH) in a 10 years old child

Type C — Low Voltage in a 6 years old child.

Seven patients which were categorized under type A exhibited the typical pattern of right ventricular hypertrophy (RVH) with strain and with possible asymmetric septal hypertrophy (ASH). Four patients under type B showed the electrocardiographic changes resembling the combined ventricular hypertrophy (CVH). The electrocardiogram included under the changes of low voltage i.e. the complex

in all leads was less than 6mm.

Discussion

The present study of electrocardiographic abnormalities in the Duchenne dystrophic patients show ed a typical ECG pattern without any cardiac symptoms. This has previously been reported by. several investigators (Gilroy et aL, 1963; Skyring and Mckusick, 1961; Wahi, 1963; Perloff et aL, 1966). The findings of sinus tachycardia, electrical axis deviation, shortening of P.R interval. tall R waves in leads V1 & V2 and deep Q waves most commonly in lead V5, V6 (and also in some limb leads) were also consistent with the previous reports (Zatuchm et al., 1951; Wahi, 1963).

The presence of deep Q waves are usually considered as the manifestation of replacement fibrosis of cardiac muscle by connective tissue and fat. Presence of deep Q waves in leads V5 and V6 along with tall R waves in V1 and V2 may represent asymmetric septal hypertrophy (ASH), but Slucka (1968), failed to demonstrate ASH in autopsies of dystrophic patients. However, recently, Chaudhary (1982) reported typical ASH changes in the ECG of one Duchenne patient. The presence of typical changes of ASH in six Duchenne patients in this series may suggest the obstructive cardiomyopathy, however, a confirmation on the basis of autopsy studies is required before reaching a conclusion. Another interesting feature observed in the present study was the presence of low voltage ECG changes in a severely affected child. These low voltage changes were indicative cf widespread myocardial damage, which seem to be proportional to the severity of the progression of the disease. +

The uniform electrocardiographic pattern independent of the duration of the disease in the present study suggests that the dystrophic process involves the myocardium from the very beginning of the disease. In most of the patients studies, the ECG changes were typical of RVH with strain, whereas in few cases the changes were of CVH with ASH and strain pattern. These distinct ECG findings suggest that scalar electrocardiograms could prove to be a sensitive index of dystrophic heart disease in the Duchenne group of muscular dystrophy.

Acknowledgements

Our thanks are due to Dr. A. Samad of the National Institute of Cardiovascular Diseases, Karachi, for his critical review of the manuscript and kind suggestions.

References

1. Chaudhary, MA. (1982) ASH in Pseudo-Hypertrophic muscular dystrophy (Abstract). Pak. Heart J., 14 : 24.
2. Emery, A.E.W (1972) Abnormalities of the electrocardiogram in hereditary myopathies. J. Med. Genet., 9:8.
3. Frankle, K.A. and Rosser, R.J. (1976) The pathology of the heart in progressive muscular dystrophy; epimyocardial fibrosis. Hum. Pathol., 7:375.
4. Gilroy, J., Cahalan, J.L., Berman, R. and Newman, (1963) Cardiac and pulmonary complications in Duchenne's progressive muscular dystrophy. Circulation, 27:484.
5. Hunter, S. (1980) The heart in muscular dystrophy. Br.Med Bull 36 ;133.
6. Ishikawa, K., Shirato, C., Yotsukura, M. Ishihara, T., Tamura, T. and Inoue, M. (1982) Sequential changes in high frequency notches on QRS complexes in progressive muscular dystrophy of the Duchenne Type; a 5 years follow-up study. J. Electrocardiol., 15:23.
7. Kovick, R.B., Fogelman, A.M., Abassi, A.S., Peter, J.B. and Pearce, M.L. (1975) Echocardiographic evaluation of posterior or left ventricular wall motion in muscular dystrophy. Circulation, 52:447.

8. Lane, R.J., Gardner-Medwin, D. and Roses, A.D.(1980) Electrocardiographic abnormalities in carriers of Duchenne muscular dystrophy. Neurology, 30:497 (1975).
9. Lukasik, E. Electrocardiographic studies in female carriers of Duchenne muscular dystrophy. J. Neurol., 209 ;279.
10. Nadas, A.S. Pediatric cardiology. Philadelphia, Saunders 1958.
11. Perloff, J.K., DeLeon, A.C. D. (1966) The cardiomyopathy dystrophy. Circulation, 33:625.
12. Perloff, J.K., Roberts, W.C., DeLeon, A.C. Jr., and O'Doherty, D. (1967) The distinctive electrocardiogram of Duchenne's progressive muscular dystrophy; an electrocardiographic pathologic correlative study. Am. J. Med., 42: 179.
13. Sanyal, K.S. and Johnson, W.W. (1982) Cardiac conduction abnormalities in children with progressive muscular dystrophy; electrocardiographic features and morphologic correlates. Circulation, 66:853.
14. Skyring, A. and Mckusick, V.A. (1961) Clinical, genetics and electrocardiographic studies in childhood muscular dystrophy. Am.J.Med. Sci., 242:5 34.
15. Slucka, C. (1968) The electrocardiogram in Duchenne Progressive muscular dystrophy. Circulation, 38:933.
16. Wahi, P.L. (1963) Cardiac changes in myopathy. Am.heart J., 66: 749.
17. Walton, J.N. and D. Gardner-Medwin, D. Progressive muscular dystrophy and the myotonic disorders, in disorders of voluntary muscle. Edited by John Nicolas, 3rd ed. EdinburghWalton. Churchill Livingston, 1974, p.561.
18. Weisenfeld, S. and Messinger, W.J. (1952) Cardiac involvement in progressive muscular dystrophy. Am. Heart J., 43: 170.
19. Zieglier, R.F. Electrocardiographic studies in normal infants and children. Illinois, Thomas, 1951.
20. Zatuchni, J., Aegerter, E.E., Moithan, L. and Shuman, C.R. (1951) The heart in progressive muscular dystrophy. Circulation, 3: 846.