Prognostic Factors Affecting Duchenne Muscular Dystrophy in Children University Hospital, Assiut University, Egypt: a cross-sectional study

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Abstract

Background& aim: Duchenne muscular dystrophy (DMD) is considered one of the most serious diseases affecting children. The disease is associated with reduced life expectancy. The current research aimed to assess possible risk factors that affect the outcome.

Results: Over one year, 82 male patients with DMD were enrolled in the study. The mean age of those patients was 13.07 years old. The majority of them (79.3%) came from rural areas. It was found that 45 (54.9%) patients were on steroid therapy. Of those patients, 44 (53.7%) were able to walk. Normal mental and language development occurred in the majority of them. It was found that patients below 13 years had better muscle tone and activity of the weight-bearing joint. Also, patients on steroid therapy had better muscle tone and activity of the weight-bearing joint. Based on the current study, older age, steroid therapy, physiotherapy, mental development, socio-economic status, presence of cardio-respiratory complications, and creatine kinase level are the predictors of the outcome of DMD.

Conclusion: DMD is a serious disease and should be managed by a multidisciplinary team. Compliance with therapy and physiotherapy has a significant effect on the outcome. Future multi-center studies with large sample size are warranted.

Keywords: Duchenne muscular dystrophy, steroid, physiotherapy, cardiomyopathy.
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Introduction:

Duchenne muscular dystrophy (DMD) is a condition primarily affecting children, with a life expectancy of less than 30 years. However, improvements in home ventilator care and spinal surgery pushed the average mortality age into the mid to late 30s [1].

Generally, DMD still has an unfavorable prognosis. However, this does not mean

that DMD is an untreatable disease. The unique medical treatment available is steroid therapy, which appears to prolong walking capacity by at least two years [2].

Many factors affect those patients and can lead to early loss of ambulation. Cardiomyopathy is one of these factors. Cardiac abnormalities become noticeable early in the course of the disease; by 18 years old, most will have developed cardiomyopathy [3].

Studies about prognostic factors of DMD in our locality are lacking, so the current work was designed to determine the prognostic factors affecting ambulation in DMD and factors that can delay the age of ambulation loss.

Methods:

Study setting& design

A cross-sectional study was conducted at the departments of Neurology, Cardiology, and Intensive Care Unit of University Children's Hospital from May 2019 to May 2020.

Study population

Eighty-two male patients in the age group 10-18 who were previously diagnosed with DMD according to:

- Typical clinical picture: (history of delayed walking, waddling gait, toe walking or frequent falling, bilateral symmetrical muscle).
- Shooting serum creatine phosphokinase (CPK) level is 50-300 times greater than normal levels [4].
- Electromyography (EMG): it allows the detection of myogenic changes even at the subclinical impairment stage. At a further stage- when compensating mechanisms were no longer possible - a dramatic decrease in amplitude, amplitude size, and density was observed [5].

Female patients with muscular dystrophy and those with autoimmune diseases or malignancies were excluded from the study.

Sample size calculation

It was calculated using open-source Open Epi info, version 3.1, considering the mean difference between the ambulatory and non-ambulatory groups regarding muscle strength of the lower limb (26±3.8 versus 23±5.7 respectively) according to Yilmaz et al. [6], with 95% confidence interval and 80% power of the test. The total required sample size was 82 patients.

Technical design:

The data collected in the questionnaire included: socio-demographic: name, age, residence, family size, family history of DMD, gestational age, birth weight of the child, and mental and motor development.

Clinical evaluation

Anthropometric measurement: weight, length, or height according to the position. Muscle strength ranges between no, trace, poor, fair, good, and normal activity [7].

Muscle tone is evaluated by passive manipulation of the limbs individually. The degree of resistance is determined to

be less than normal (hypotonic), normal, or more than normal (hypertonic). The last may be referred to as spasticity [7]. The range of movement of weight-bearing joints was assessed.

Besides the full physical examination, the following data were gathered; steroid use, vitamin use, CPK level, presence or absence of cardiomyopathy based on echocardiography, and myopathic changes based on EMG.

***The North Star Ambulatory Assessment (NSAA)*[8].**

NSAA is a 17-item scale that grades the performance of various functional skills on a scale from 0 (unable), 1 (completes independently but with modification), and 2 (completed without compensation). This scale is ordinal, with a total of 34 degrees. The maximum score indicates a fully independent function.

Statistical analysis

Data management: entry, revision, cleaning, and recoding were done using SPSS for Windows program (version 20.0, SPSS, Chicago, IL, USA). Descriptive analysis was done; frequency and percentage for qualitative variables and median and range for NSS after testing for normality as it was not normally distributed. Chi-square or Fisher's exact test was used to test the difference in frequencies/percentages between qualitative variables in both groups.

Mann Whitney / Kruskal-Wallis test was used to compare the NSS between/among groups.

Linear regression analysis was done to determine factors affecting the NSS. Statistical significance level was considered when $p \leq 0.05$ in all tests**.**

Result

The mean age of patients was 13.07 ± 2.32 years. Most of the cases (79.3%) were rural residents. About three-quarters of the mothers were housewives, and 43.9 of the fathers were farmers. About a fourth of the cases had a family history of DMD.

Unfortunately, about half of the cases (51.2%) were going to school; only 33.3% were going on foot. Regarding steroid therapy, 54.9% were currently using steroids; another 36.6% previously used steroids, while 8.5% never used steroids.

Among current steroid users, the majority (82.2%) were on interrupted use, and 13.3% were on every other day regimen. As regards dietary supplements, about 70.7% were receiving dietary supplements, while 29.3% stopped dietary supplements, and the mean duration of the dietary supplement was 4.14+1.68 years. More than half of the cases (53.7%) were on physiotherapy.

Cardiac examination shows that 35.4% had cardiomegaly, and 17.1% had heart failure. Only half of the patients with cardiac problems received medications; angiotensin-converting enzyme inhibitors (ACIs) for all treated and B-Blockers (35.0%). Fig (1) shows that more than half of the patients (53.7%) are still walking, and 40.2% use wheelchairs. Among the walking patients, 93.2% have walking difficulty, 27.3% only can up and/ or downstairs, whereas 9.8% had used a

bicycle for physiotherapy. Fig (2) shows that most cases (85.4%) had normal mental development, whereas 74.4% had delayed motor development. Ambulation is significantly higher among the younger age group, patients with employed mothers, those with normal mental development, those on current steroid use, with normal respiratory examination, without cardiomyopathy, and those on cardiac treatment. Patients with current use of steroids had significantly better muscle power and tone in both upper and lower limbs and better range of movement in weight-bearing joints. NSS is significantly higher among the younger age group, sons of working mothers, small family size, those with normal cardiac and respiratory examination, with normal mental and motor development, and among those under physiotherapy, dietary supplementation, and current steroid use. Figure (3) shows a significant negative correlation between body mass index (BMI) and NSS. Fig. (4) shows a significant negative correlation between CPK level at the time of diagnosis and NSS. Linear regression analysis showed that the significant factors increasing the NSS score were the use of steroids, normal motor developmental history (CI is 1.1-9.9 and 3.3-12.9 respectively), whereas NSS was significantly decreasing with older age and higher BMI (CI is -2.6 to -0.9 and -1.1to -0.3 respectively).

Discussion

The present study revealed that patients with DMD deteriorate with aging, as seen from the mean of NSS. This result is in agreement with Scott et al. [9]. They demonstrated a progressive decline of muscle strength with age and a good correlation between muscle strength and motor ability. This explains the higher percentage of ambulation reported among the younger age group.

Our study revealed increased respiratory complications and cardiac involvement with increasing age. This is in agreement with Bushby et al. [10]. Eagle et al. [11] reported that the cause of death in DMD patients is typically respiratory and/or cardiac complications.

Corticosteroid therapy is helpful for patients with DMD, as documented by mean NSS. This finding is conformed to Bushby et al. [12]. Biggar et al. [13] and Manzur et al. [14] demonstrated that glucocorticoid therapy had been shown to have multiple benefits, including delaying loss of muscle function, loss of ambulation, and improving mobility.

Also, Silva et al. [15] confirmed that steroid therapy prolongs walking capacity by at least two years. Moreover, Gloss et al. [16] revealed that glucocorticoid therapy is considered the standard of care for Duchenne.

Our study confirmed that the interrupted use regimen of steroids was the most commonly used, while the least common is the continuous use regimen. This is in agreement with Pradhan et al.[17] who reported that a continuous regimen is not preferred due to its complications.

Physiotherapy is essential to prolong ambulation, as documented by the mean NSS. This is in line with Bushby et al. [10]

reported that physiotherapy is imperative for alleviating muscle atrophy, skeletal deformities, and motor function deterioration. Also, Vasanth et al. [18] reported that physiotherapy also helps in preventing contracture, which can prolong independent locomotion.

Mazzon et al. [19] demonstrated that the importance of physiotherapy is attributed to the importance of normal range of movement of weight bearing joint in ambulation, and strength loss as in hip extension and ankle dorsiflexion is considered the primary predictor of loss of ambulation in DMD leading to progressively reduced walking capacity.

Patients with delayed mental development had a poor prognosis, as shown by mean NSS. This is in line with Mochizuki et al. [20], who reported that 38% of DMD patients exhibited slight mental retardation. This group began walking in the later period, lost walking ability earlier, and died earlier.

Patients with a history of delayed motor development have a poor prognosis compared to those with normal motor development. Also, Hinton et al. [23] reported that early developmental delay implicates an underlying central nervous system component to DMD, which predicts poor prognosis.

A positive family history of DMD had no significant effect on the prognosis of DMD patients.

This may be due to the low flow of patients with a family history of DMD due to the high economic and emotional burden in transporting the affected children for follow-up visits, especially after feeling no hope in treatment, particularly after doing their best with the previously affected child.

As regards cardiomyopathy, patients with cardiac medication have a good prognosis and better walking capacity. In agreement with our results, Towbin [21] reported that early therapeutic intervention would benefit DMD patients with LV dysfunction even if asymptomatic.

A significant negative correlation was revealed between body mass BMI and NSS. In agreement with our results, Bushby et al. [12] and Hankard et al. [22] reported that obesity can aggravate difficulties in locomotion and increases the burden on already weakened muscles.

There was a significant negative correlation between CPK level and NSS at the time of diagnosis. This is due to an increased level of serum CPK is the hallmark of muscle damage and its marked elevation at the start of the disease indicates severe damage to the muscle. Moreover, its decrease with age indicates no more muscle bulk for a breakdown, as seen in figure (2). This result is in agreement with Zats et al. [24].

The prognosis of DMD was better among sons of employed mothers and small-sized families. This can be explained by the employee's mother's increase in the family's income, which is needed to pay for the patient's care regarding their follow-up visits, physiotherapy sessions, and good compliance to treatment of this lifelong chronic disease. Also, all this care

is more affordable among small-sized families.

In line with our results, Galbraith et al. [25] and Okumura et al. [26] reported that children living in higher socio-economic status families were able to have family care replaced by professional care (chest physiotherapy and psychic support), which those children in lower socio-economic status families were unable to have.

Conclusion and recommendations:

Based on the current study, factors negatively affecting the prognosis of DMD were age and BMI, whereas steroid therapy and normal motor development history were positively affecting the prognosis. So, DMD cases must be treated in specialized centers with multidisciplinary teams to manage BMI and ensure compliance with treatment regimens, including steroid therapy to prolong ambulation status and prevent or at least delay complete disability. More attention must be paid to children who showed delayed motor development in early life to be managed early and properly.

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Tables

Table (1): Socio-demographic characteristics and family history of DMD cases

	No. (82)	**%**
Age: (years)		
10 - < 13	42	51.2
≥ 13	40	48.8
Mean ± SD (Range)	13.07 ± 2.32 (10.0-18.0)	
Residence:		
Rural	65	79.3
Urban	17	20.7
Occupation of mother:		
Housewife	61	74.4
Employee	21	25.6
Occupation of father:		
Farmer	36	43.9
Employee	30	36.6
Skilled worker	8	9.8
Free business	8	9.8
Family history of DMD	21	25.6
Relation:	(N = 21)	
Brother	17	81.0 %
Uncle	7	33.3 %

Table (2): Therapeutic history and Schooling and hobbies among DMD cases

	No. (82)	**%**
Patients who go to school	42	51.2
How does the patient go to school?	(N = 42)	
Walking	14	33.3
By car/ bus	28	66.7
Patients who have any favorite sport	3	3.7
Physiotherapy:	44	53.7
Steroid therapy:		
Current use	45	54.9
Previous use	30	36.6
Never use	7	8.5
Duration of steroid use:		

Mean ± SD (Range)	4.40 ± 1.54 years (2.0 - 8.0)	
Regimen of current steroid use:	(N= 45)	
Interrupted	37	82.2
Every other day	6	13.3
Continuous	2	4.4
dietary supplement (L- carnitine) :	58	70.7
Duration of dietary supplement: Mean ± SD (Range)	4.14 ± 1.68 years (1.0-7.0)	

Table (3): Cardiac and respiratory function among DMD cases

	No. (82)	**%**
Cardiac examination*		
Normal	42	51.2
Cardiomegaly	29	35.4
Heart failure	14	17.1
Cardiac medication:	(N= 40)	
Yes	20	50.0
No	20	50.0
Type of cardiac medication*:	(N= 20)	
ACIs	20	100.0
ACIs & B-blockers	7	35.0
Respiratory examination:		
Normal	48	58.5
Pneumonia	23	28.0
Respiratory failure	9	11.0
Pleural effusion	2	2.4

***Multiple answers**

Table (4): Socio-demographic and clinical factors affecting the ambulation status among DMD cases

	Walking				**X^2**	**P-value**
	Yes (n= 44)		**No (n= 38)**			
	No.	**%**	**No.**	**%**		
Age:					17.58	**<0.001**
10-<13Y	32	76.2	10	23.8		

≥13Y	12	30	28	70		
Residence:					1.05	0.305
Rural	33	50.8	32	49.2		
Urban	11	64.7	6	35.3		
Occupation of mother:					8.46	**0.004**
Housewife	27	44.3	34	55.7		
Employee	17	81.0	4	19.0		
Family size:					7.43	**0.024**
4 – 5	18	69.2	8	30.8		
6 – 7	23	53.5	20	46.5		
> 7	3	23.1	10	76.9		
Family history of DMD:					2.75	0.097
Yes	8	38.1	13	61.9		
No	36	59.0	25	41.0		
Cardiomyopathy:					35.58	**<0.001**
Yes	8	20.0	32	80.0		
No	36	85.7	6	14.3		
Cardiac medication:	(N= 8)		(N= 32)		Fisher	**0.044***
Yes	7	87.5	13	40.6		
No	1	12.5	19	59.4		
Respiratory examination:					35.44	**<0.001**
Normal	39	81.3	9	18.8		
Abnormal	5	14.7	29	85.3		
Mental development:					7.74	**0.005**
Normal	42	60.0	28	40.0		
Delayed	2	16.7	10	83.3		
Current steroid use:					43.70	0.001
Yes	39	86.7	5	13.5		
No	6	13.3	32	86.5		

Table (5): Muscle activity and range of movement of the weight-bearing joints among DMD cases according to current steroid use

	Current Steroid use				X^2	P-value
	Yes (n= 45)		No (n= 37)			
	No.	%	No.	%		
Muscle power:					31.16	0.001*
Upper limb:						
No/ Trace activity	1	2.2	19	51.4		
Poor/ Fair	15	33.3	12	32.4		
Good/ Normal	29	64.4	6	16.2		
Lower limb:					34.50	0.001*
No/ Trace activity	2	4.4	24	64.9		
Poor/ Fair	33	73.3	11	29.7		
Good/ Normal	10	22.2	2	5.4		
Muscle tone:					46.76	0.001*
Upper limb:						
Average	42	93.3	7	18.9		
Hypo	3	6.7	30	81.1		
Lower limb:					46.76	0.001*
Average	42	93.3	7	18.9		
Hypo	3	6.7	30	81.1		
Range of movement of Weight-bearing joint:					40.63	0.001*
Hip:						
Full	40	88.9	7	18.9		
Limited	5	11.1	30	81.1		
Knee:					40.63	0.001*
Full	40	88.9	7	18.9		
Limited	5	11.1	30	81.1		
Ankle:					9.73	0.002*
Full	19	42.2	4	10.8		
Limited	26	57.8	33	89.2		

Table (6) North Star score according to socio-demographic data and some clinical factors in DMD cases

Variable	Median (Range)	P-value
Age:		
10-<13Y	18.0 (1.0-34.0)	**0.001**
≥13Y	2.0 (0.0-27.0)	
Residence:		
Rural	8.0 (0.0-34.0)	0.142
Urban	17.0 (1.0-34.0)	
Occupation of mother:		
Housewife	4.5 (0.0-34.0)	**0.008**
Employee	17.5 (0.0-34.0)	
Family size:		
4 – 5	16.5 (1.0-34.0)	**0.02**
6 – 7	7.0 (0.0-34.0)	
> 7	1.0 (0.0-32.0)	
Family history of DMD		
Yes	3.0 (0.0-29.0)	0.245
No	14.0 (0.0-34.0)	
Cardiac examination:		
Normal	18.0 (1.0-34.0)	**0.001**
Abnormal	1.0 (0.0-17.0)	
Respiratory examination:		
Normal	17.0 (0.0-34.0)	**0.001**
Abnormal	1.0 (0.0-34.0)	
Mental development:		
Normal	14.0 (0.0-34.0)	**0.015**
Delayed	1.0 (0.0-30.0)	
Motor development:		
Normal	19.0 (7.0-34.0)	**0.02**
Delayed	7.0 (0.0-34.0)	
Physiotherapy:		
Yes	17.0 (1.0-34.0)	**0.002**
No	2.0 (0.0-34.0)	
Dietary supplement:		**0.001**

Yes	16.0 (0.0-34.0)	
No	1.0 (0.0-13.0)	
Current use of steroids:		
Yes	17.0 (1.0-34.0)	**0.001**
No	1.0 (0.0-34.0)	
Walking:		
Yes	18.0 (1.0-34.0)	**<0.001**
No	2.0 (0.0-27.0)	

Figures:

Fig (1): Ambulation status among DMD cases

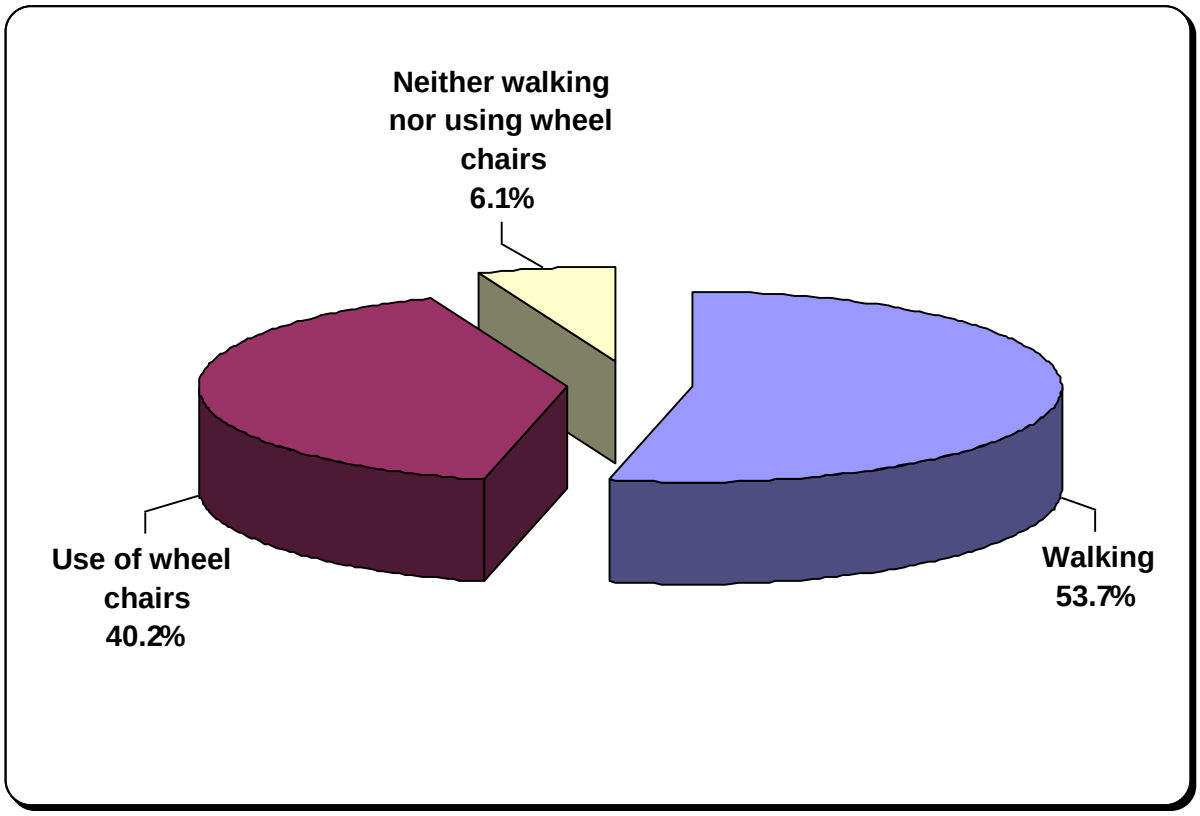
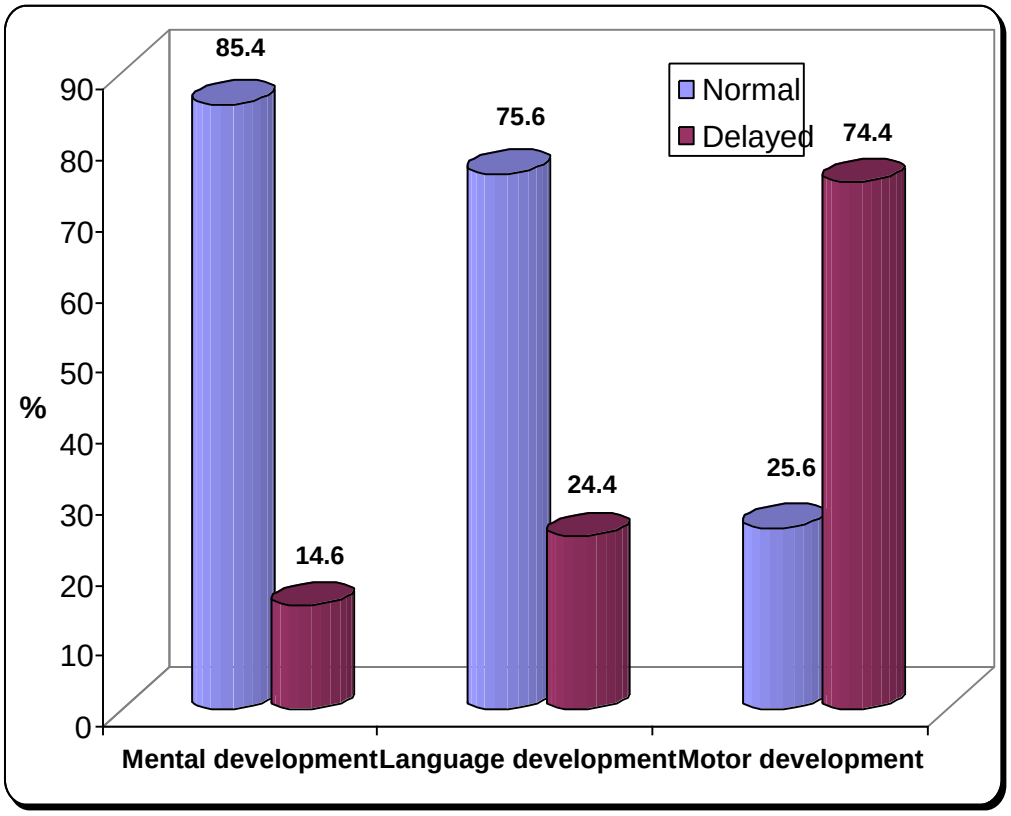


Fig (2) Motor and mental development among DMD cases



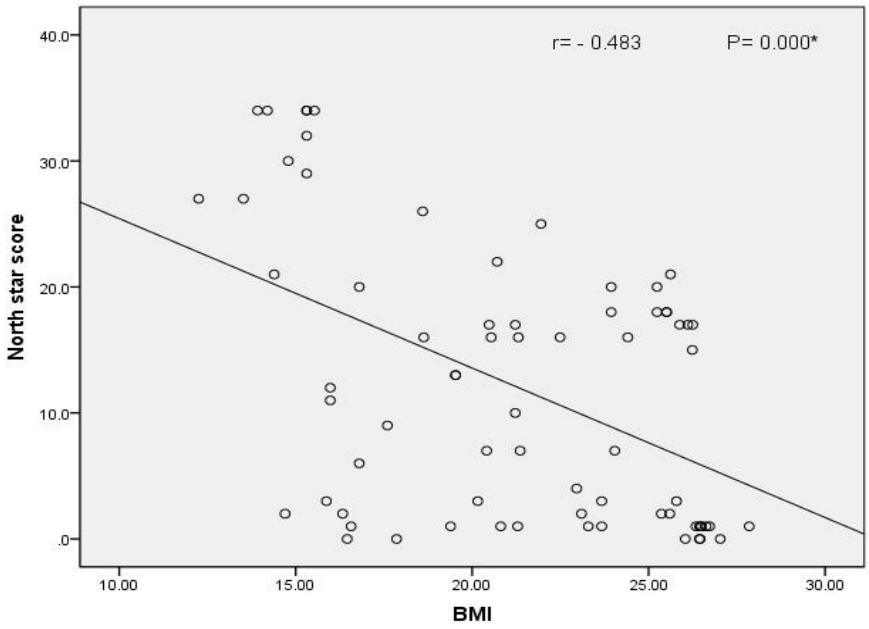


Fig (3): Correlation between BMI and North Star score among DMD cases

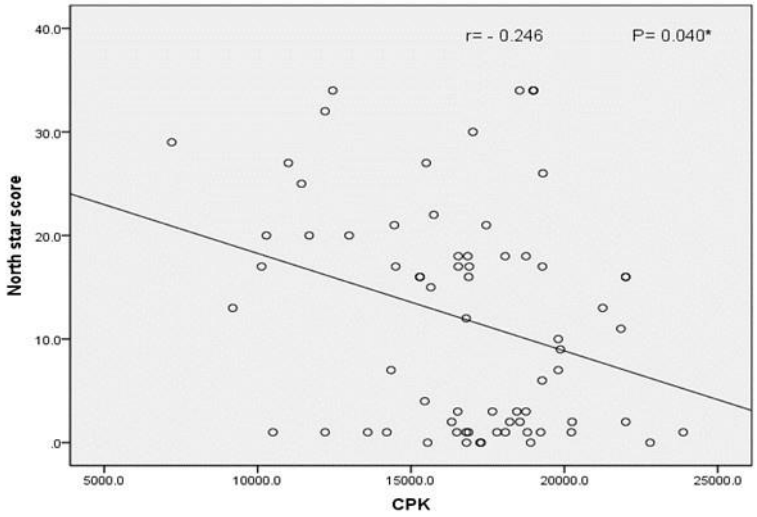


Fig (4) Correlation between CPK and North Star score of DMD cases