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Research Article

The Effect of Corticosteroids in the Treatment of Muscle Weakness in Patients with Duchenne Muscular Dystrophy at Buali Hospital In 2015

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ABSTRACT
Introduction: Duchenne muscular dystrophy (Duchenne Muscular Dystrophy, DMD) is the second most common fatal genetic disease in humans and is the most severe form of muscular dystrophy. This disease infects muscle and is associated with destruction of muscle fibers. Given that even if the diagnosis is still no effective treatment for this disease and there is nocertain cure for Duchenne disease, some patients take treatment that aims to slow the progression of the disease. Given that the effect of corticosteroid therapy in the treatment of DMD is not still completely proven and had not been a study about it in our country, In this study, rate of muscle weakness in patients with Duchenne muscular dystrophy treated with corticosteroids and placebo were compared. Materials and Methods: In this study, this was conducted as a randomized double-blind clinical trial, 30 patients with Duchenne muscular dystrophy were enrolled in 1393 at buali Hospital and The improvement in muscle strength in both groups were compared for a month. Finally The data whichobtained, were analyzed with statistical software spss19 . Results: The results showed that muscle strength before intervention was similar in both groups (P> 0.05)؛ But after the intervention, muscle strength was significantly higher in the corticosteroid group than in the placebo group. (P=0.025). 9 patients (60%) in the corticosteroid group and two patients (13.3 percent) in the placebo group had improved that showed a statistically significant difference (P = 0.008).
Conclusion: In sum, based on the results of this study suggest that corticosteroids in the treatment of muscle weakness in patients with Duchenne muscular dystrophy are effective and their use in the treatment of these patients is recommended.

Keywords: corticosteroids, muscle weakness, Duchenne muscular dystrophy

[I] INTRODUCTION:
Duchenne muscular dystrophy (Duchenne Muscular Dystrophy, DMD) is the second most common fatal genetic disease and the most severe form of muscular dystrophy in humans (1).The incidence of this type of myopathy is 1 in every 3,500 live male births and its prevalence is estimated 1 per 100 live births (2-3).However, reports of girls suffering mild disease have also been reported (1).Most babies had normal birth by natural evolution and the skills of sitting, standing, is normal or with a slight delay.A small number of patients may also have loose tone and weakness at first day of birth. Disease Symptoms typically appear between the ages of 5-3 years and include the inability to climb stairs, fall is frequent and walking on tip toes.Weakness often symmetric pattern and starting from the hip and shoulder girdle, which also involved the respiratory muscles and distal limbs.Death is often due to respiratory failure occurred inthe age about 20 years, but in those who survive by ventilator had greater life expectancy, often the cause of death was heart failure (4-6).Given that even if found and diagnosis, yet there is no effective treatment

for this disease and so far there is no cure for DMD gene therapy is still in the preliminary stages and research (1,3).Some patients take treatments aimed at slowing the progression of disease. Although steroids temporarily, but only pharmacological approach that progress in slowing loss of muscle strength and motor ability in patients with DMD plays a role. Steroids also possible to delay the respiratory failure, cardiac involvement and scoliosis. Nevertheless but so far their use as a standard treatment for DMD is still unproven (7-10). In a study of Parreira et all in 2010 in Brazil, muscle strength and motor function in patients with Duchenne muscular dystrophy who received steroids from time to time compared with each other.In this study, 90 patients with Duchenne muscular dystrophy which one to seven years of steroids received from the Medical Research Council Scale (MRC) and the Hammersmith motor ability score were compared and it was found that the progression of the disease with steroids is delayed (7). Given that the effect of corticosteroid therapy in the treatment of DMD is not yet fully proven and it's also in our study had not been, in this study, muscle weakness in patients with Duchenne muscular dystrophy treated with corticosteroids and placebo were compared.

[II] MATERIALS AND METHODS:

This interventional study to double-blind randomized clinical trial controlled clinical trial Daubl-blind randomized on 30 patients with Duchenne muscular dystrophy in 1393 was admitted to Buali hospital in 1393.The study was conducted after reviewing the inclusion criteria and obtains informed consent from all patients. The final diagnosis in patients with Duchenne muscular dystrophy patients and informed consent, patients were enrolled and treated patients who have undergone surgical procedures or other treatment, were excluded. All the information that was taken and the name of the patient in this study remained confidential. Studied patients were divided randomly into two equal groups of 15.A group treated with prednisolone 75/0 mg per kg body weight for a month. The other group treated with placebo. Glorified by the Sobhan pharmaceutical company with the same view and similar taste to prednisolone were prepared. Drug and placebo encoded by the pharmaceutical company and participants in study such as theexecutor of plan and also patients, did not know anything about the type of drug. After completion of treatment,
muscle strength and associated symptoms in patients in both groups were analyzed by the following methods.

A: 6-minute walk test for that patient during walking muscle strength was evaluated by a doctor.

B: Mayomtryktst: a device for determining the quality of eccentric muscle strength.

Variables studied included age during the study, gender, age ofbeginning treatment, response to treatment, muscle strength and course of the disease in people in a questionnaire prepared is entered. Finally, after gathering the information required from all participants, we attempted to analyze the data in this field we used the statistical software SPSS version 13. To review the existing statistical relationship Kayaskvar test, Fisher exact and T-test was used and the significance level was five-hundredths of a point.

[III] RESULT:

All patients in both groups were male. Table 1

		Gender	
		Male	Total
Group	Corticosteroid	15	15
		100.0%	100.0%
	Placebo	15	15
		100.0%	100.0%
Total		30	30
		100.0%	100.0%

Table 1 . Distribution of sex of studying patients

9 patients (60%) in the corticosteroid group and two patients (13.3 percent) in the placebo group had improved that showed a statistically significant difference (P = 0.008). Table 2

		Improvement		
		Pos	Neg	Total
Group	Corticosteroid	9	6	15
		60.0%	40.0%	100.0%
	Placebo	2	13	15
		13.3%	86.7%	100.0%
Total		11	19	30
		36.7%	63.3%	100.0%

Table 2. Distribution of recovery in studying patients

Muscle strength after the intervention was significantly higher in the corticosteroid group than in the placebo group (P = 0.025). The mean age, age of beginning the therapy and duration of therapy in patients studied in two groups and muscle strength before intervention was similar in both groups (P> 0.05).

[IV] DISCUSSION AND CONCLUSION:

The results showed that muscle strength before intervention in two groups were matched; but after the intervention muscle strength was significantly higher in the corticosteroid group than in the placebo group. 9 patients (60%) in the corticosteroid group and two patients (13.3 percent) in the placebo group were improved, showed no statistically significant differences. In a study of Parreira et all in 2010 in Brazil, muscle strength and motor function in patients with Duchenne muscular dystrophy who received steroids from time to time compared with each other. In this study, 90 patients with Duchenne muscular dystrophy who received steroids from one to seven years the Medical Research Council Scale (MRC) and the Hammersmith motor ability score were compared. And it was observed that with steroid use is delayed disease progression (7) that the results obtained in our study are consistent with the prescribed short-term corticosteroid. In a study by Ricotti et all was conducted in 2013 in England, glucocorticoids effect was evaluated in the treatment of Duchenne muscular dystrophyand the benefits and side effects of prednisolone discontinuous and daily therapy in 360 patients werecompared.The averageLoss of ambulation in 12 years discontinuous therapy and 14/5 years daily therapy. Moderate to severe side effects, like Cushingoid appearance, behavioral effects and hypertension was more common in daily treatment. BMI also had higher daily treatment. Dwarf was more sever in daily treatment (11). However, in our study due to short-term administration of prednisolone no side effect was observed. In a study Shimomura and colleagues in 2011 in Japan, long-term effects of prednisolone in the treatment of Duchenne muscular dystrophy in 14 treated patients and 15 controls who lack the ability to walk were compared and no significant difference in losing ambulation ability in the treatment group and the control observed and in this study prednisolone did not prolong the study period of ambulation (12).However, in our studyshort-termadministration of prednisoloneshowed goodefficacyin compared to placebo. In a study by Rahman et al in 2001 in Bangladesh, 19 patients with Duchenne muscular dystrophy in a randomized clinical trialchecked out and the effects of prednisolone were evaluated.To 10 patients at 0.75 mg / kg prednisone was given for 6 months, 8 patients were treated with vitamins and it was observed that muscle strength significantly better in the group was treated with prednisone (13)which in turn are consistent with the results obtained in our study. In sum, based on the results of this study and comparison with other studies in this field has been said that Corticosteroids in the treatment of muscle weakness in patients with Duchenne

muscular dystrophy are effective and their use in the treatment of these patients is recommended.But in the end recommendedthat again, more research in this area to confirm these findings obtained in this study with larger sample sizes to be done.

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