EXERCISE MANAGEMENT FOR PEOPLE WITH MCARDLE DISEASE : A PILOT CASE STUDY



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Background

McArdle disease is a rare metabolic condition characterized by deficiency of enzyme myophosphorylase. This is a case study of a 61 year old male who has been diagnosed with the condition 5 years ago. The cardinal signs and symptoms are inclusive of activity intolerance, early fatigue and cramping within few minutes of initiating an activity that leads to muscle damage, rhabdomyolysis and potential renal failure.

Methods

A six weeks protocol included an initial baseline functional assessment for the client prior to exercise session. The exercise session was inclusive of a 7-10 minutes of warm up, 20-30 minutes of cardiovascular and resistance training followed by a 10 minutes of cool down phase. There were two supervised sessions per week. After 10 sessions of exercise protocol functional assessment was repeated and results noted. The client was allowed proper rest periods during the exercise session. For dietary intervention the client took guidance from sports nutritionist. During the session client's heart rate was monitored and rate of exercise was graded on Rate of perceived exertion scale.

Exertional cramps leads patients to shun from exercise. Some studies have reported aerobic exercises are an effective means of improving physical activity in people suffering from this disease whereas others suggest that resistance training helps to bring about better clinical outcomes.

Currently the effectiveness of low carbohydrate ketogenic diet has also been found effective for this disease condition. Hence, the client in our study has already been going through ketogenic diet planned by the nutritionist.

Apart from the underlying issue, the client also suffers from white coat syndrome.

Purpose

Through this pilot study we have tried to investigate on effectiveness of combination of aerobic and resistance

Results and Discussion

Post intervention the functional assessment revealed a significant improvement in agility, muscular strength, muscular endurance, balance, flexibility of upper body and aerobic capacity of the client. Client also reduced weight and waist girth. Overall improvement resulted in gaining confidence for the client.

The exercise induced response is due to the main energy substrate phosphocreatine that has no dependence on muscle glycogen deposits. The rest intervals allow for the resynthesis of phosphocreatine before it gets utilized again.

training for the client suffering from McArdle's Disease.



Weight (kg)	

 0
 10
 20
 30
 40
 50
 60
 70
 80
 90
 100

	Weight (kg)	waist girth (cm)	RHR (bpm)	TCL (mmol/L)	BGL (mmol/L)	Agility	Handgrip (kgF)	muscle endurance	Arm Curl(30 sec)	back scratch (cm)	single leg balance (erro
After	69	91.5	66	7.2	5.6	3.47	32	16	16	19.5	2
Before	e 71.1	95	77	3.8	6.7	4	24	13	7	28	8

Ketogenic diet on other hand induces nutritional ketosis that helps to improve exercise tolerance and reducing the risk of muscle damage and further complications by lowering plasma creatine kinase.

Conclusion

We put forward an exercise regime to increase physical activity and quality of life in people suffering from this condition. Low muscle mass and strength levels lead to comprised cardiometabolic phenotype. Hence, exercise-induced hypertrophy is an attainable goal in McArdle's patients.

References

1. Reason, S.L., Cadzow, R., Jegier, B.J., & Wakelin, A. (2018). Centre of Expertise: Comprehensive care for patients with McArdle Disease. Journal Rare Disorders:

After Before

Scope for Further Research

Further we will try an investigate the effectiveness of 12 weeks individualised rehab program followed by a period of no exercise intervention. Also adding a component of protein or creatine supplements may or may not enhance muscle mass gain.

Diagnostic & Therapy, 4(1:4). DOI: 10.21767/2380-7245.100174.
Munguia-Izquierdo, D., Santalla, A., & Lucia, A. (2014). Cardiorespiratory fitness, physical activity, and quality of life in patients with McArdle disease. Med. Sci. Sports Exercise, 47(4), 799-808. DOI: 10.1249/MSS.0000000000000458
Kitaoka, Y. (2014). McArdle disease and exercise physiology. Journal of Biology, 3, 157-166. DOI: 10.3390/biology3010157
Baller,R.G., Wyrick, P., Taivassalo, T., & Vissing, J. (2006). Aerobic conditioning: An effective therapy in McArdle's disease. Ann Neurol, 59(6), 922-928. DOI: 10.1002/ana.20881
Santalla, A., Munguia-Izquierdo, D., Brea-Alejo, L., Pagola-Aldazabal, I., Diez-Bermejo, J., ..., & Lucia, A. (2014). Feasibility of resistance training in adult McArdle patients: Clinical outcomes and muscle strength and mass benefits. Frontiers in Aging Neuroscience, 6(334). DOI: 10.3389/fnagi.2014.00334