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## What are the causes of muscular dystrophy in the elderly?

1. 0 Introduction
Muscular dystrophy refers to a group of degenerative disorders that are hereditary and progressive which result in the weakening of the skeletal muscles. There is a variety of muscular dystrophy which differ in terms of onset, degree of severity, penetrance and pattern of muscles that are affected. The main types of muscular dystrophy include: facioscapulohumeral, congenital, myotonic, Duchenne, limb girdle and oculopharnygeal and distal. Some of these disorders manifest during the formative years of life but some only manifest later on. This therefore implies that muscular dystrophy is a group of degenerative disorders that affects people of all ages; infants and the elderly included. However, the scope of this paper shall be limited to muscular dystrophy in the elderly.
Muscular dystrophy in the elderly is as a result of both the aging process and genetic factors which result in the production of molecules that are highly reactive and free radicals. It has been found that the free radicals and highly reactive molecules trigger a vicious cycle in which a set of group protein known as ryanodine receptors leak calcium into the cell. Given that there are various types of muscular dystrophy affecting the elderly, this report shall seek to outline the causes, intervention measures; the future forms of treatment by reviewing previously conducted studies on muscular dystrophy.
1. 1 Search strategy
The search strategy entailed finding the following key words: muscular dystrophy, causes, risk factors elderly, treatment and outcomes. The scope of the search was limited to peer-reviewed materials only. The search was conducted on the following online databases: NCBI, Pubmed, Ageing Journal and Orphanet Journal of Rare Diseases.

1. 2 Inclusion and exclusion criteria
There was no restriction to the date of the publication of the journals used in the review. This was done in order to obtain a broader perspective into the area of interest. The study had to revolve around muscular dystrophy in the elderly population in line with the objectives of the study. In addition, this was also done to get an overview of the progress made in research as far as the disorder is concerned. Studies on children and adolescents were excluded from the search. The methods used in the studies were explicitly outlined. The result of these studies was clearly stated. The aims and objectives were in line with the objectives of this report. Most of the articles were qualitative given that they provided an in depth understanding into the causes and were inclined towards explaining the phenomenon at hand.
Six of the articles used were qualitative journal articles, two were quantitative journal articles, three were web based qualitative articles and finally, one was a quantitative government report.
2. 0 Data synthesis and analysis
2. 1 Population groups
Muscular dystrophy disorders affect the elderly. This has been attributed to the increased production of free radicals and highly reactive molecules which results in calcium leaks in ryanodine receptors which ultimately results in weakening of the muscles. The weakening and withering of muscles is referred to as sarcopennia and is common among individuals aged between 40 and 75 years. The onset of muscular dystrophy is not only attributed to age but is also attributed to genetic factors .
Some of the forms of muscular dystrophy that are associated with ageing include: oculopharnygeal muscular dystrophy and myotonic dystrophy. Oculopharnygeal muscular dystrophy is a late onset progressive disease which has been found to affect individuals aged 45 years and above which is characterized by atrophy of the tongue, dysphagia, ptosis and weakening of the proximal lower limbs. In one study, it was found that the average age at which individuals developed ptosis was 48 years while dysphagia 50 years old. Late onset myotonic dystrophy affects individuals from the third and fourth decades of life.
2. 2 Prevalence
Studies in the United States have shown that the disease is more prevalent among individuals of French Canadian descent . The prevalence of oculopharnygeal muscular disorder in France is 1: 100, 000 while in Canada particularly among French Canadians, prevalence stands at 1: 1000. These rates are applicable for the autosomal dominant form of OCPD. The autosomal recessive form prevalence rate is 1: 10, 000 in France. The occurrence of the disease is not only limited to French Canadians but it affects other nationalities too.
It is estimated that oculopharnygeal muscular dystrophy affects about 30 nations. A number of Spanish Americans in Texas and California have also been found to have oculopharnygeal muscular dystrophy as per the results of one study. In one study of 12 family members, it was found that 4 of members were affected while two were asymptomatic carriers. The authors noted that there are no records in Asia showing the occurrence of the disease which they attributed to the masking of the disease by the late onset and other conditions such as myasthenia gravis
2. 3 Causes
There are two causes that result in muscular dystrophy among the elderly: ageing and genetics. Ageing results in the production of free radicals and molecules that are highly reactive. These molecules interrupt the normal functioning of ryanodine receptors by resulting in the leaking of calcium. The leaked calcium interferes with the mitochondria which are responsible for energy production within the cells. As a result of this, there is increased leakage of calcium hence less calcium is available for the contraction of muscle fibres. Gradually, the muscle fibres loss their contractile ability and weaken. There are several forms of muscular dystrophy that are caused by genetic defects. Myotonic dystrophy type 2 is a progressive, inheritable disease that is characterized by muscle weakness, pain and stiffness. It affects both young adults from the age of 20 years and older ones who are aged 50 years and above. It is caused by an “ unstable 4 nucleotide repeat expansion on chromosome 3.”
Oculopharnygeal muscular dystrophy can be passed on from parent to offspring in tow fashions: autosomal recessive and autosomal dominant. The risk of inheriting the OCPD gene is 50% for the autosomal dominant gene. On the other hand, the risk of the offspring inheriting the OCPD autosomal recessive gene is less than 1% because in such instances the offspring are obligate heterozygotes of a mutant allele of OPMD. Facioscapulohumeral muscular dystrophy mostly affects individuals below the age of 20years but a case has been reported of an individual aged 77 years being diagnosed with the disease. Facioscapulohumeral muscular dystrophy is the result of a deletion in the DZ4 segment of chromosome 4.
2. 4 Interventions
For OCPD, the intervention measures are pegged down to the type of symptom. Ptosis can be corrected using surgery specifically two forms of blepharoplasty which are: frontal eyelid suspension and resection of levator palpebrae aponeurosis. Surgical correction is necessary for individuals with severe forms of dysphagia as a result of the OCPD. Annual flu vaccinations ought to be administered to affected individuals in addition to minimising the isolation and cutting the foods given to the affected individuals into smaller bits.
For muscular dystrophy as a result of ageing, a drug known as s107 has successfully undergone clinical trials. In a study involving mice that were 24 months old, it was found that the mice that were administered with the drug had muscles that were 50% stronger than those that received the control. There are suggestions that corticosteroids might prove to be useful in slowing down the effects of facioscapulohumeral disease. Creatine which is often used by atheletes to enhance their performance has also been found to be useful in slowing down the symptoms of the disease. Severe forms of myotonic muscular dystrophy could be treated using phenytoin and quinine which are meant to relieve the symptoms of the disease. Rehabilitation is also necessary for patients suffering from any form of muscular dystrophy.
3. 0 Summary of findings
There is a general consensus among scholars that muscular dystrophy is as a result of ageing and genetic factors. Until recently, the exact role of ageing in muscular dystrophy was not fully understood. However with the advances made in molecular medicine, it has now emerged that as an individual continues to age, the production of antioxidants and highly reactive molecules increases. These molecules cause calcium to leak out of the ryanodine receptors. The leaked calcium ends up in the cell where it results in the gradual detriment of the mitochondria. More calcium leaks out of the ryanodine receptors hence is unavailable for the contraction of the muscle fibres. As a result, the muscles gradually loose their ability to contract and weaken. Genetic factors also contribute to muscle dystrophy in old age. Oculopharnygeal muscular dystrophy is the result of the mutation of the gene PABN1 while facioscapulohumeral muscular dystrophy is the result of a deletion in the DZ4 section of chromosome 4. Myotonic muscular dystrophy is the result of repetition of nucleotide on chromosome three.
There is no known cure for the genetically induced causes of muscular dystrophies. However, corticosteroids are often prescribed in order to relieve the symptoms of facioscapulomuscular dystrophy. Surgery may be recommended in order to relieve dysphagia and ptosis which occur as a result of oculopharnygeal muscular dystrophy. Myotonic muscular dystrophy can e treated using quinine and phenytoin although this is only necessary for severe forms of the disease .

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