

A Brief Study on Motor Neuron Disease; and its Treatment Includes Drug Delivery System; Future Aspects on Neurological Science

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Abstract: Motor neuron disease is vary hazards disease in recent time, MND (Motor Neuron Disease) basically it shows clinical presentation to ALS (Amyotrophic lateral sclerosis) considered is most neurodegenerative disease. In MND basically loss of nerves in the spine and brain is not working so it is Refferd to neurodegeneration. It is recorded as in last past year we have seen lots of changes related to patient care treatment improving and drug safety but also rapid scientific advances is there, so that rational therapies based on key pathogenic mechanisms now seem plausible. ALS have both properties in heterogeneous. it occurs 1 year to delay from first symptoms to diagnosis where as half of patient succumb within 3-4 years , although the heterogeneity suggest therapeutic study with respect to clinical study. RNA Process is also implicated and show protein degradation. Apparent clinical heterogeneity tells us that therapeutic studies should include detailed biomarker profiling, they even tell us about the genetic and clinical statistritions. The most common mutation, accounting for 10% of all Western hemispheres ALS due to this protein degradation is occurs. Major part is still not clear how these fundamental is working continuously. This pre- symptomatic study considered is new era or new initiative for neurodegenerative disorders in neurological sciences.

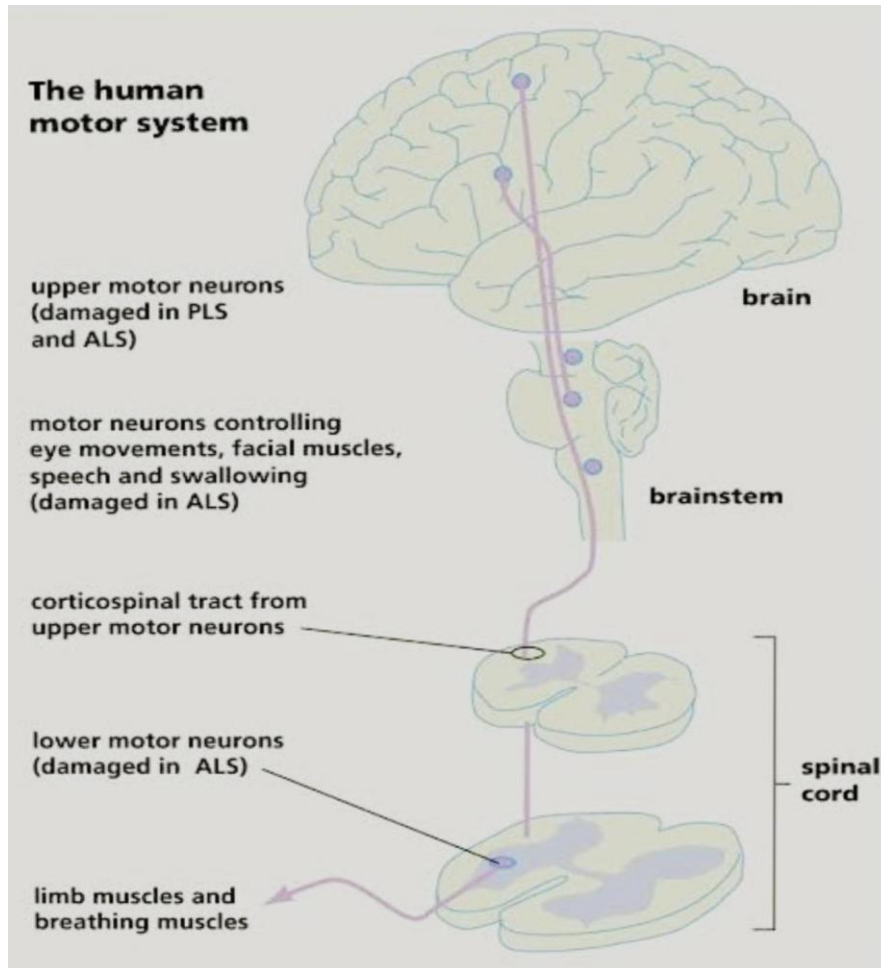
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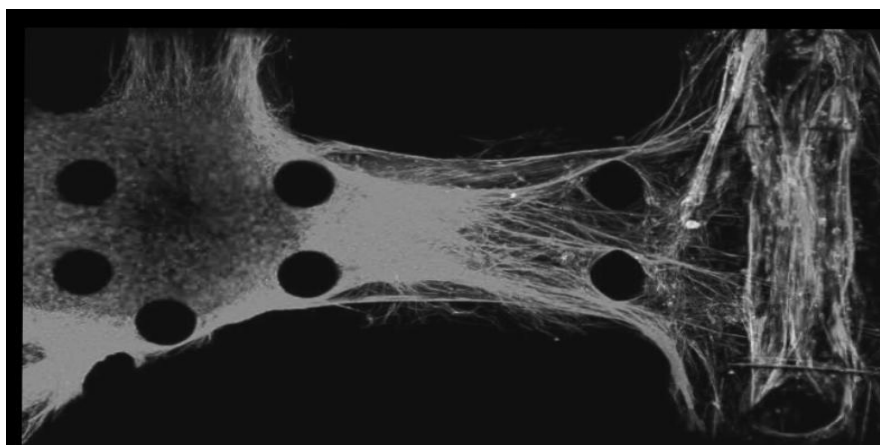
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I. Introduction

Motor neurone disease (MND) it is a group of diseases which reffers to nerves and spine or brain loss function is an adult-onset neurodegenerative disorder characterised by loss of upper motor neurons from including the Betz cells of the motor cortex), and lower motor neuron (LMNs, anterior cells of the spinal cord and brainstem nuclei (Brain). Which is shown in “Figure:1 ALS affects motor neurons in the motor cortex (UMN) and anterior horn of the spinal cord (LMN) and corticospinal tract “ and “Figure : 1.2 Drug Targeting on ALS Motor Neurons “ respectively.¹ The term MND is largely synonymous with amyotrophic lateral sclerosis (ALS), reflecting the observation that most patients demonstrate combined LMN-related loss of muscle as a result of denervation hence there is no cure or no reversible treatment for the normal Functioning neuron motor disease. ²



“Figure:1 ALS affects motor neurons in the motor cortex (UMN) and anterior horn of the spinal cord (LMN) and corticospinal tract “



“Figure: 1.2 Drug Targeting on ALS Motor Neurons “

II. Clinical Features And Diagnosis

The clinical hallmark of ALS is a progressive motor weakness without sensory disturbance. The diagnosis is not easy MND, because the main reason is it has common symptoms other conditions like multiple sclerosis (MS), or Parkinson’s disease. The primary clinical Diagnosis a general physician is referred to Neurologist those has experience in neurological disorders.

There are several parameters of neurologist perform as per patient past records are respectively :

- 1) Blood and urine tests
- 2) MRI Brain Scan

- 3) Lumbar puncture
- 4) Muscle biopsy
- 5) Electromyography (EMG)

Further, these test Neurologist must have verified patient really have MND they will monitor the patient performance before going to the final conclusion.

III. Epidemiology

ALS is considered to occur throughout the world, though knowledge is incomplete, especially in Africa, India, and China, where systematic epidemiology has not yet been carried out.³ Hotspots for the ALS-Parkinsonism-Dementia complex, described in the Japanese Kii Peninsula and on the Pacific island of Guam, are exceptional, still poorly understood in terms of etiology, and pathologically distinct. A UK population-based study found an ALS incidence of 4.6 per 120,000 population per year for women, and 4.9 per 120,000 for men, resulting in lifetime risk of 1 in 452 and 1 in 390, respectively.⁴ ALS is extremely rare below the age of 35 years, though cases linked to genetic mutations in FUS are seen in teenagers, and have a typically aggressive course.⁴ For older person risk may be increased after the age of 45 year's before a decline in incidence occurs that is unexplained.

IV. Genetics

A genetic contribution to ALS was long thought to be significant for only 5% of cases with a family history consistent with Mendelian inheritance. Incomplete gene penetrance and small family size are only two of many factors that led to an under-appreciation of the inherited contribution to ALS. For nearly two decades, only one-fifth of 'familial' cases (2% of all ALS) were accounted for genetically, through mutations in the superoxide dismutase-1 gene (SOD1). Now three-quarters of cases with a family history of either ALS or FTD are explained as autosomal dominant single gene disorders.

V. Biomarkers

There is a need for biomarkers that might reduce diagnostic delay, or improve prognostic stratification and therapeutic response assessment.²² Trials currently rely on survival as the primary endpoint, or change in the slope of the revised ALS Functional Rating Score (ALSFRS-R), both of which lack sensitivity. CSF neurofilaments, TDP-43, and neuroinflammatory molecules may reflect aspects of disease pathogenesis and progression, but need validation in large cohorts. Neurophysiology offers quantitative LMN biomarker candidates, for example, Motor Unit Number Estimation (MUNE) and electrical impedance myography. The drug to demonstrate a survival benefit in human ALS is riluzole, thought to have a broadly anti-glutamatergic mode of action. Its effects are very modest, prolonging means survival from 12 to 15 months in the clinical trial purpose. The therapeutic improvement includes those aimed at muscle function which is shown in "figure: 1.3 overview skeletons muscle Plasticity "

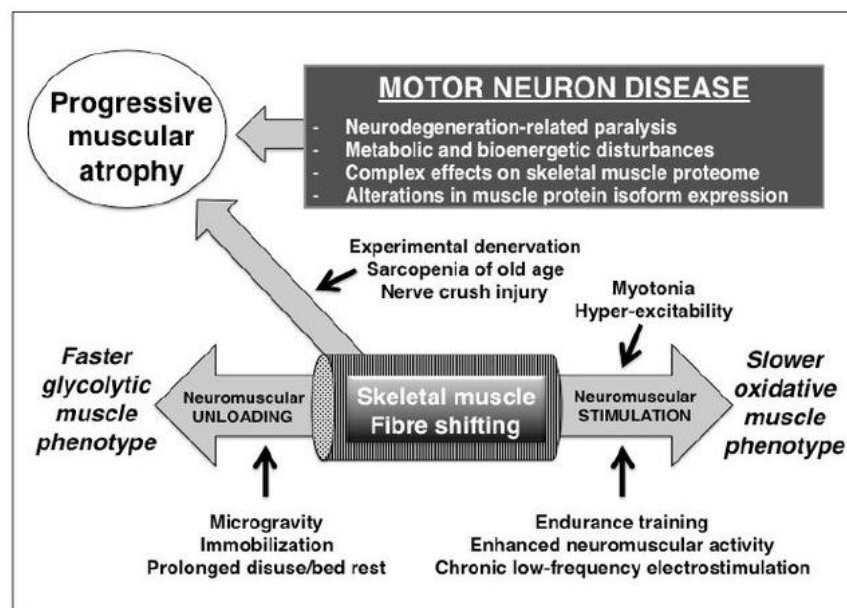


Figure 13. Overview of skeletal muscle plasticity and fi bre transitions due to physiological adaptations or pathological insults to the neuromuscular system. Motor neuron disease is characterized by progressive muscular atrophy and complex changes in the skeletal muscle proteom “

“Therapy

Treatment for drooling

This can be treated by the use of a drug named “Scopolamine”, which is even helpful in the treatment of motion sickness.

Muscle cramps and stiffness

Muscle cramps and stiffness can be treated with physical therapy and medications, such botulinum toxin (BTA) injections. This drug blocks the brain to transfer signals to stiff muscles. Baclofen (a muscle relaxer), may also reduce muscle stiffness.

Uncontrolled laughter or crying

Antidepressants, called serotonin reuptake inhibitors which control emotions and control cry.

Swallowing difficulties (dysphagia)

In these kind of patients eating and swallowing becomes difficult hence they may need a percutaneous endoscopic gastrostomy

Breathing problems

Respiratory muscles usually weaken gradually, but a sudden deterioration is possible.

Stem cell transplant for ALS treatment

More scientific study is needed for stem cell research and gene therapy for treating ALS.

Outlook

MND is usually fatal. Depending on the type, most people will not survive longer than 5 years after symptoms appear but few people live more than 10-12 years.

VI. Conclusion

There is no Permanent cure of the Patient whom suffering MND but Lots of therapy is there for maintain there body balance and mind balance such as physical, occupational and speech therapy etc. These Therapy mostly works slowly, improvement is totally depends on person it self or comfort. Scientifically two main drugs are approved by FDA (Food Drug Administration) USA. These following drug are used under NDDS; they targeted particular site for an actions like Riluzole, or Rilutek they have low amount glutamine present appears most effective in the early stages of ALS and in older individuals. Future development for ALS is doing by scientists continue research on stem cell which may be recognised in future. As the medical research is being continued, scientists understand MNDs more fully by doing research. They are working to finding new treatments.

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Conflict of interests

The authors declare that there is no conflict of interests regarding the publication of this paper.

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