

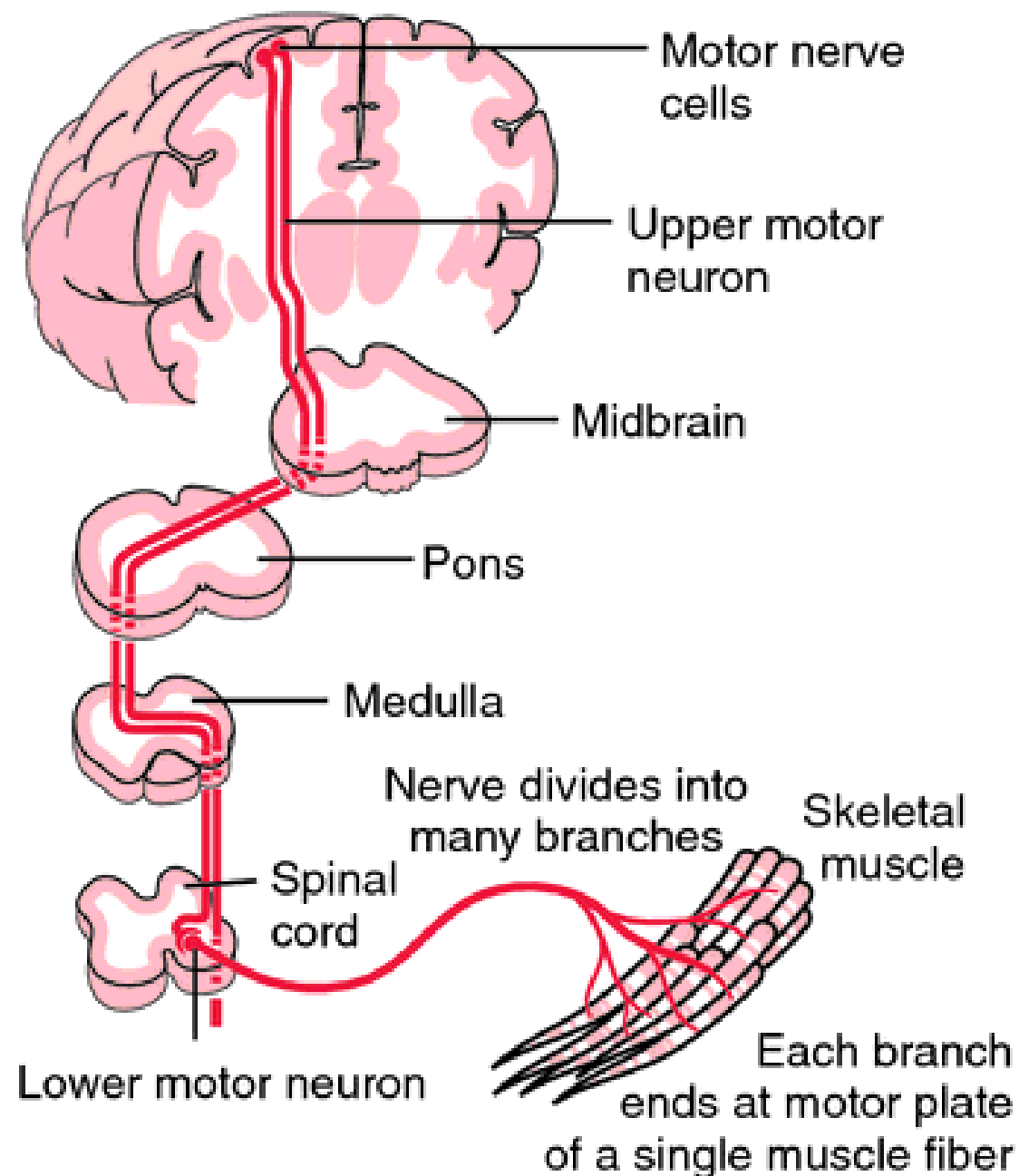


# ESCLEROSE LATERAL AMIOTRÓFICA

Neurobiologia dos Estados Patológicos  
Discentes: Helmut Kennedy, Isaac Braga e Júlia Xavier

# Esclerose Lateral Amiotrófica

- Doença neurodegenerativa progressiva que atinge os neurônios motores do córtex cerebral, tronco encefálico e medula espinal.
- 2 casos por 100.000 habitantes



**S E M I O L O G I A**



DIFICULDADE  
PARA ANDAR



DIFICULDADE  
NAS ATIVIDADES  
DIÁRIAS



TROPEÇAR  
E CAIR



ESPASMOS



FRAQUEZA  
MUSCULAR



FALA  
ARRASTADA



CAIMBRÃS  
MUSCULARES



MOVIMENTOS  
DESASTRADOS



DIFICULDADE  
EM MANTER  
A POSTURA



DIFICULDADE  
EM ENGOLIR

ISTO É *possível*



# ETIOLOGIA

**ELA**

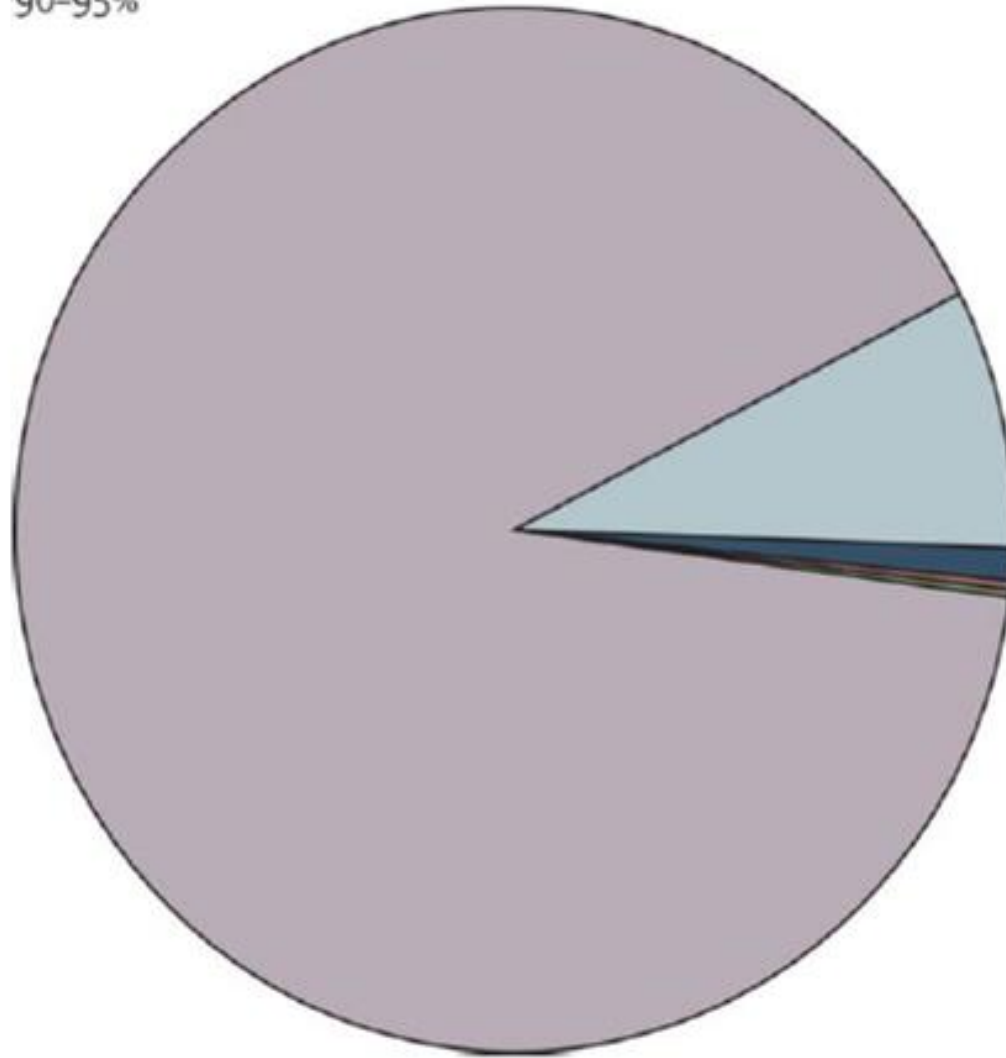


**ESPORÁDICA**

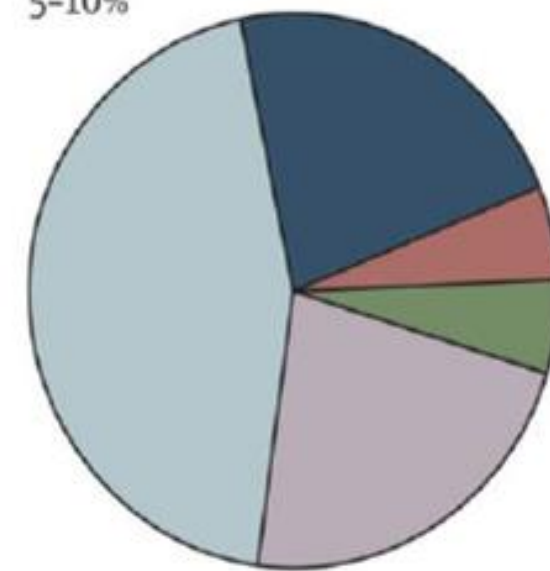


**FAMILIAR**

**Apparently sporadic ALS**  
90-95%

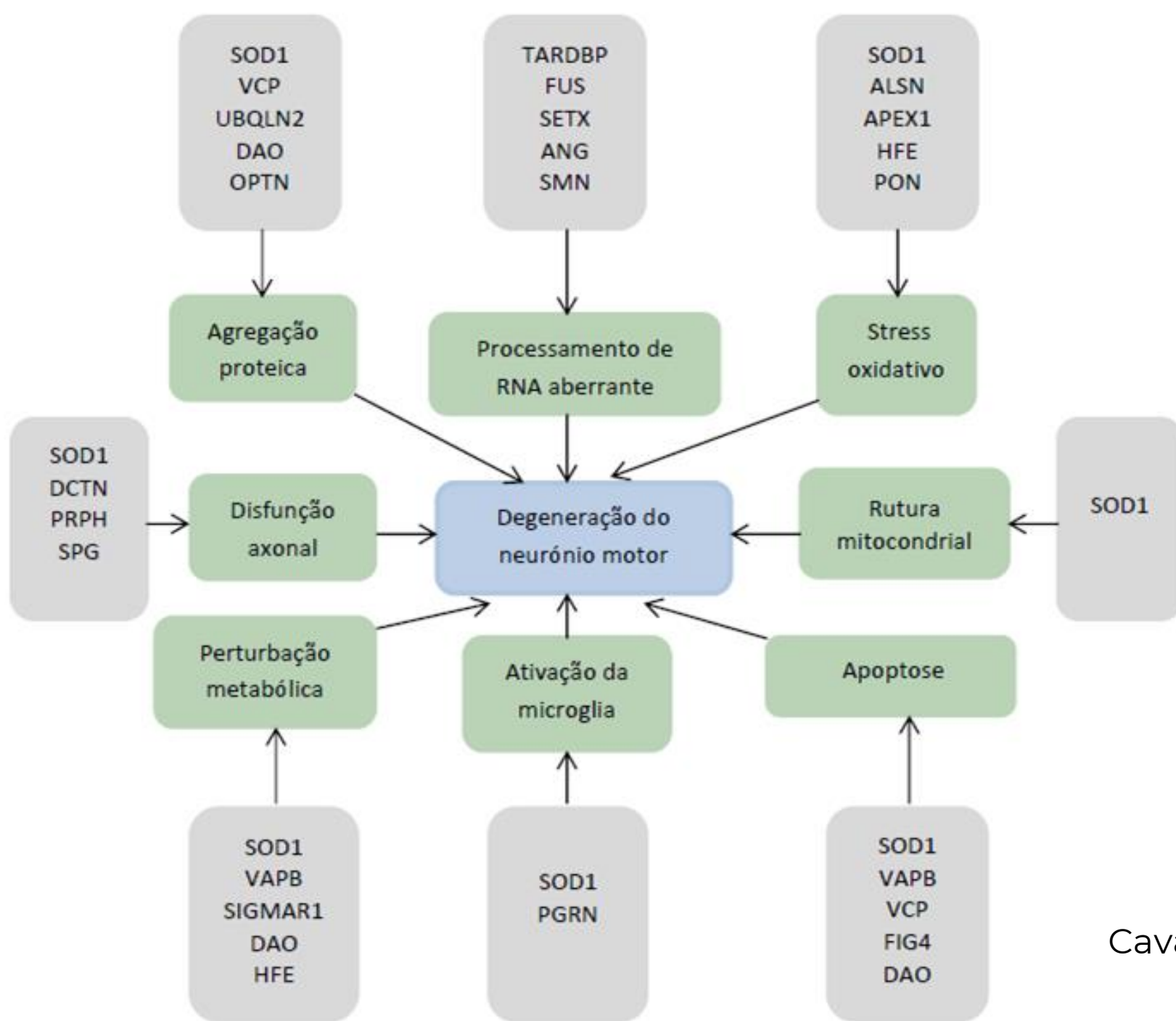


**Familial ALS**  
5-10%



**FISIOPATOLOGIA**



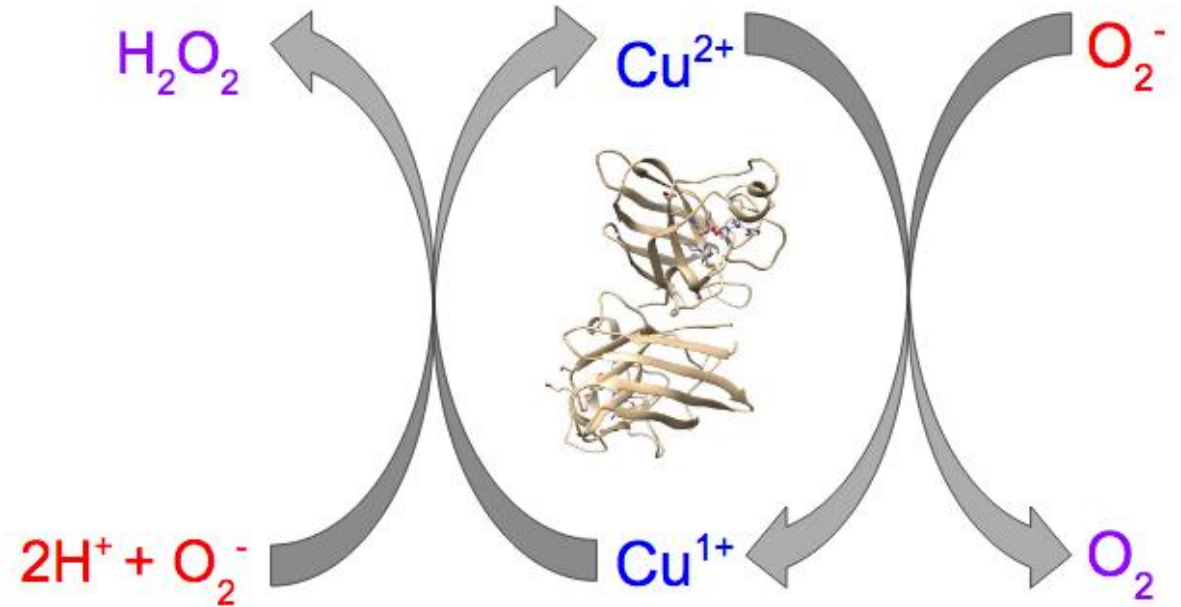


Cavaco, S.G., et al. 2016

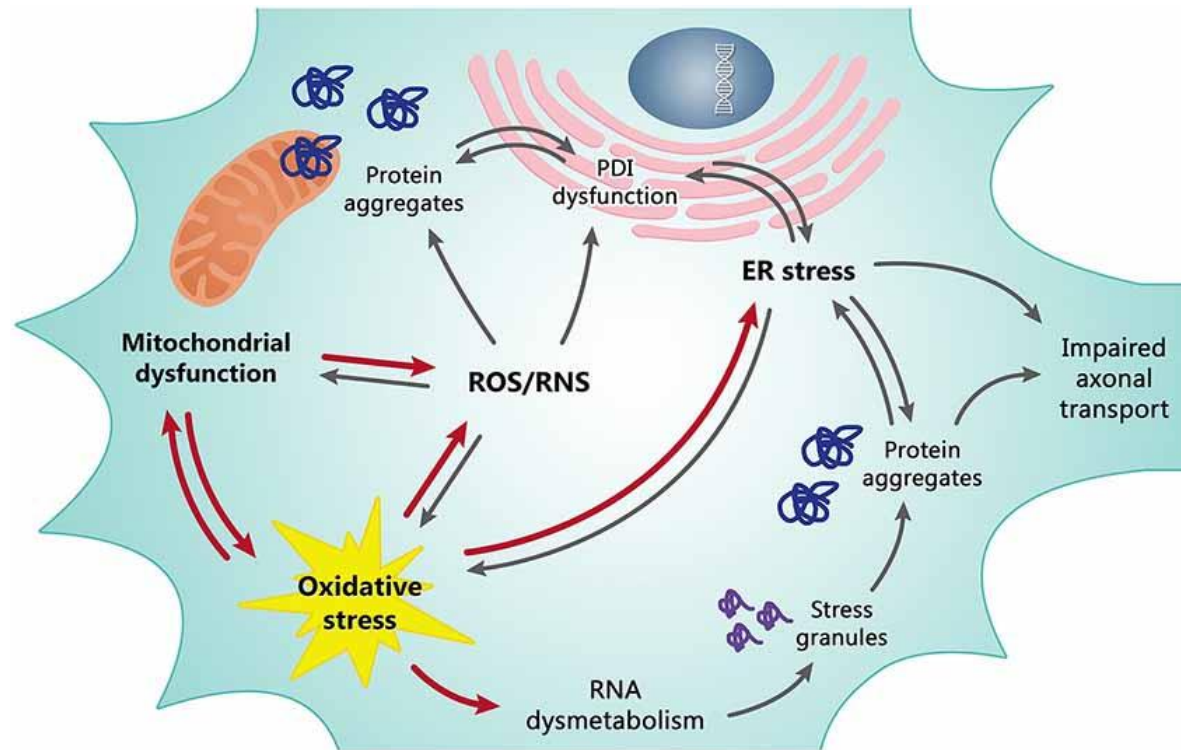
# Stress Oxidativo

SOD1

- Perda da função catalítica;
- Ganho de função - Interações aberrante.

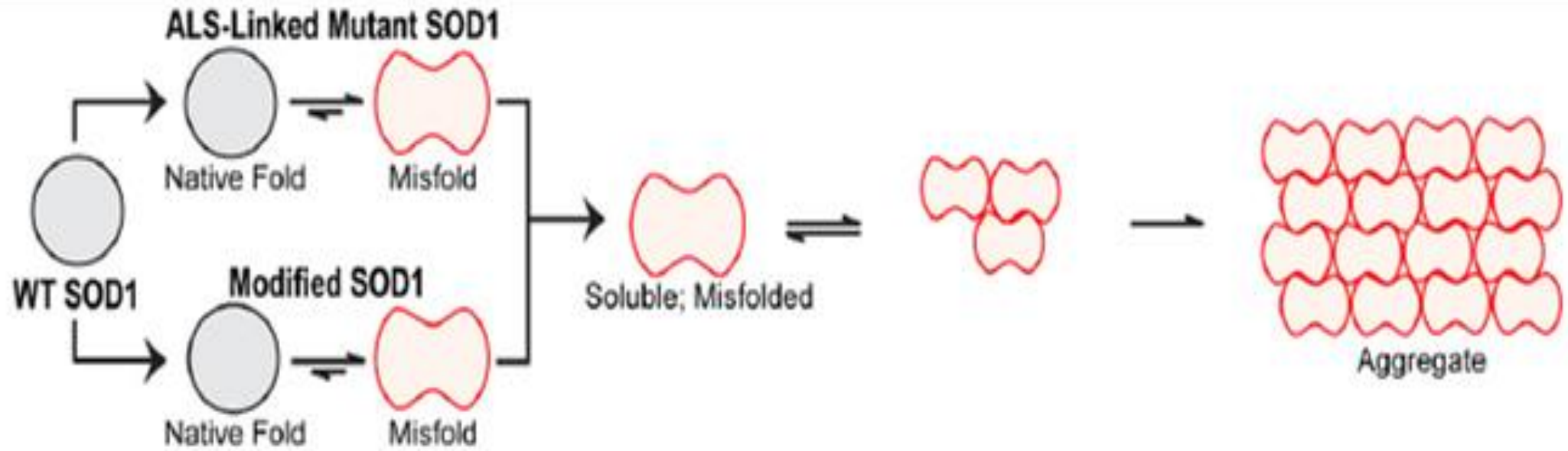


# Stress Oxidativo



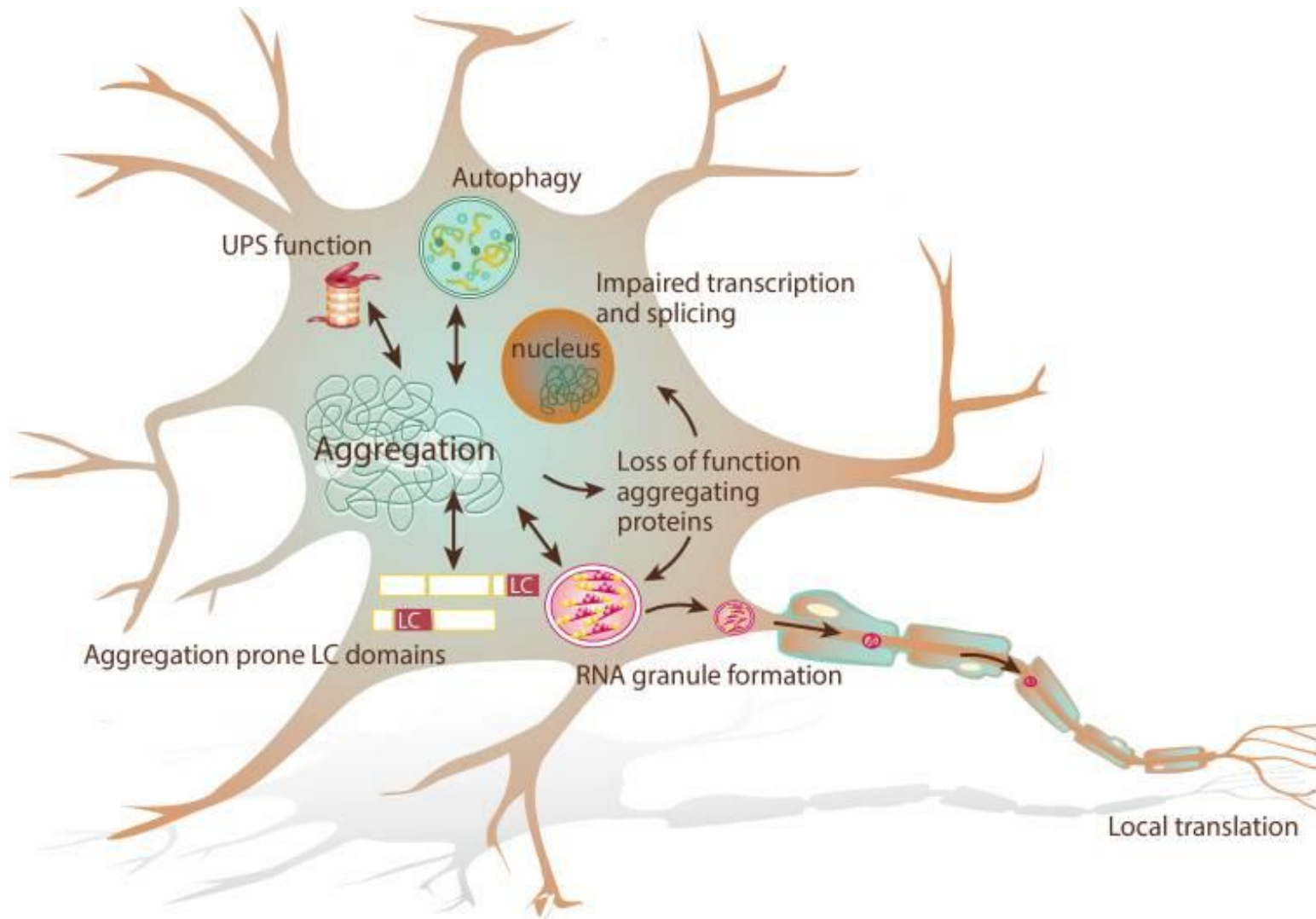
Carri et al, 2015

# Agregação Proteica



Rotunno and Bosco , 2013

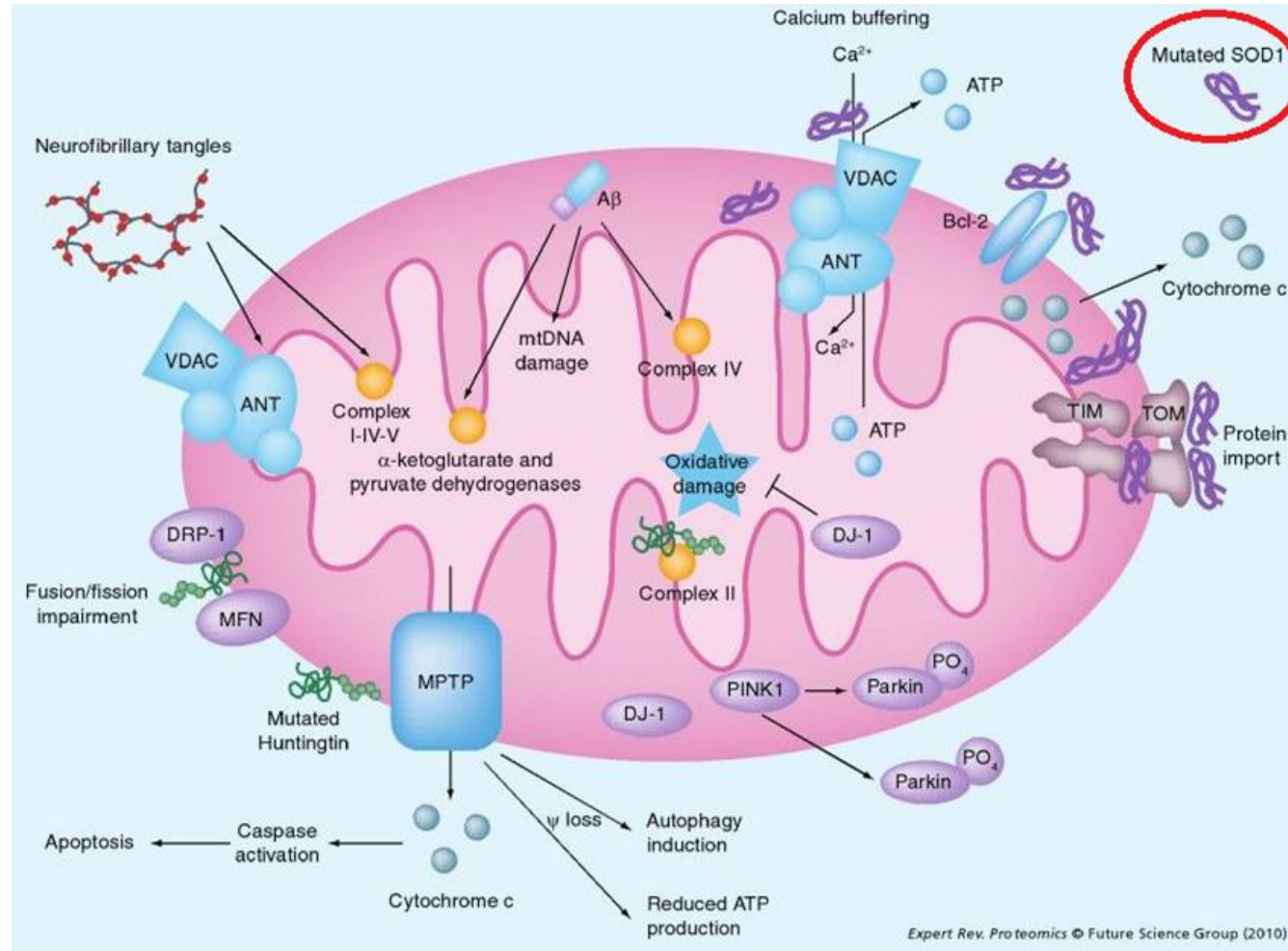
# Agregação Proteica



Blokhuis et al, 2013

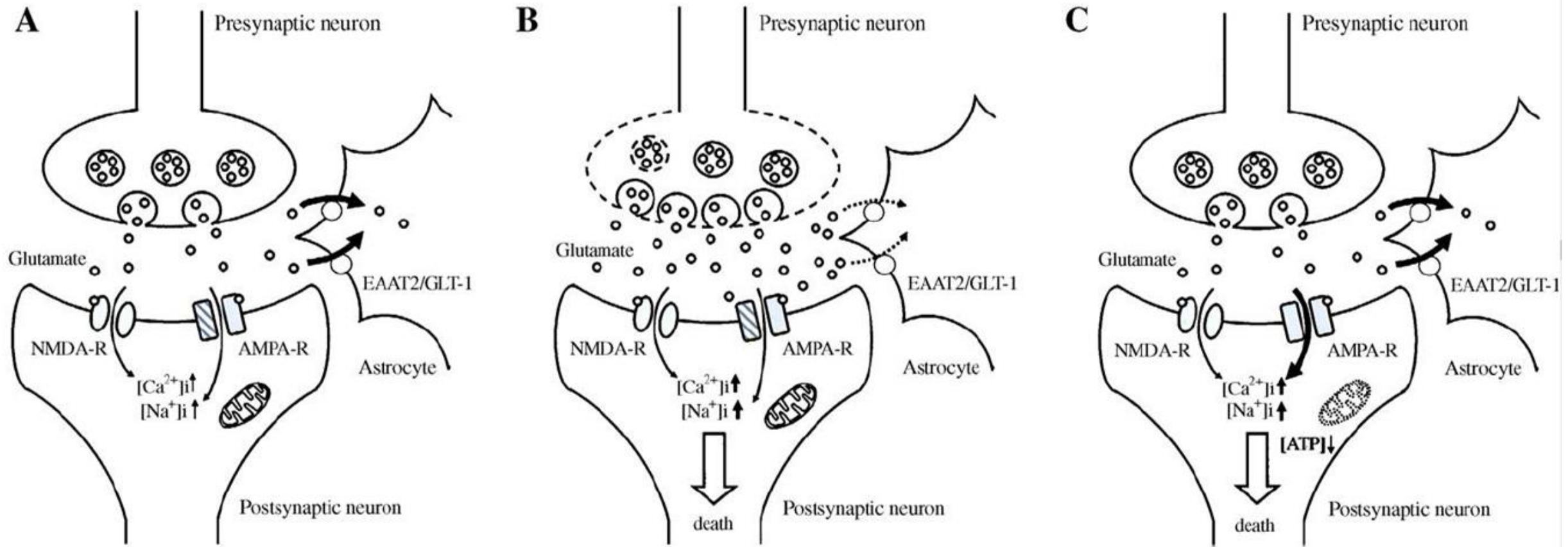


# Disfunção Mitochondrial

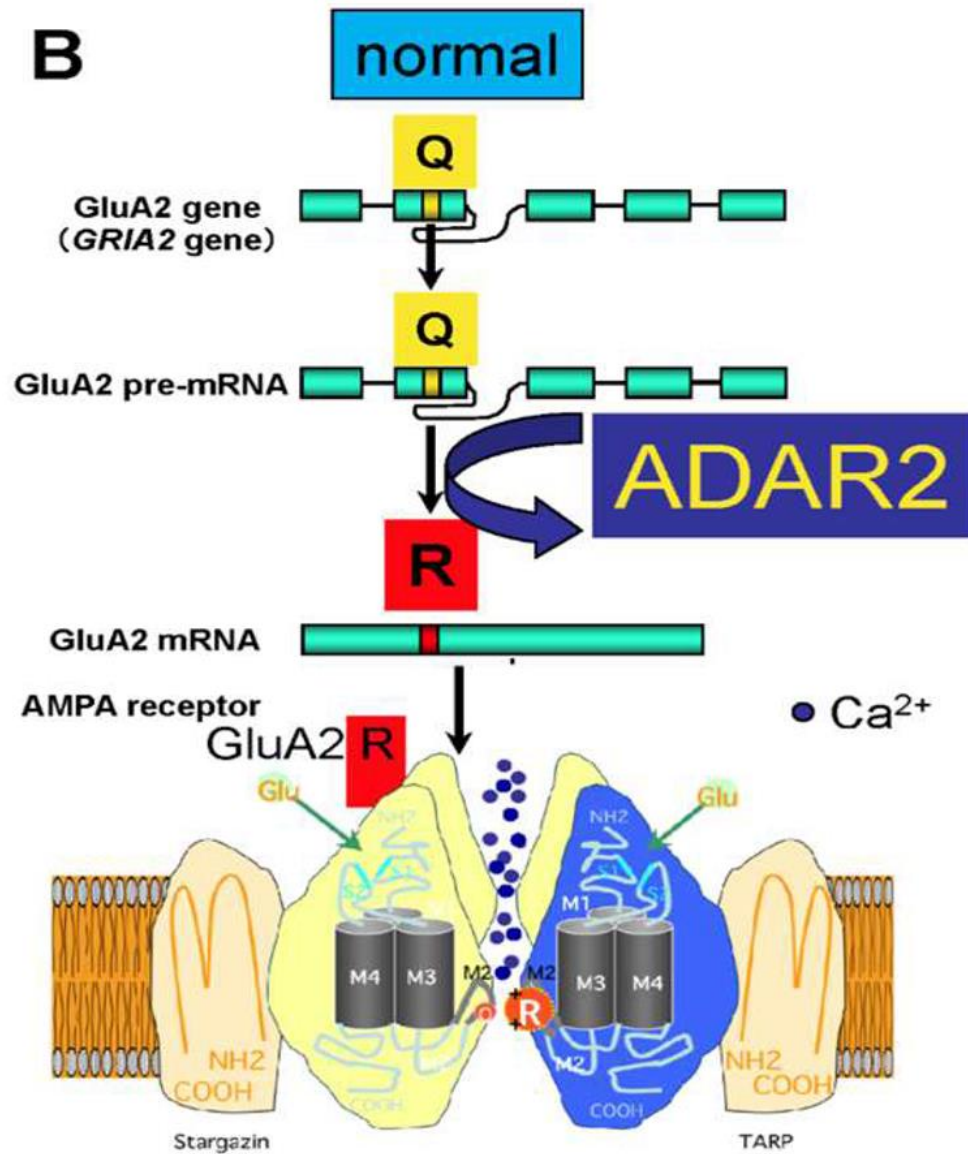
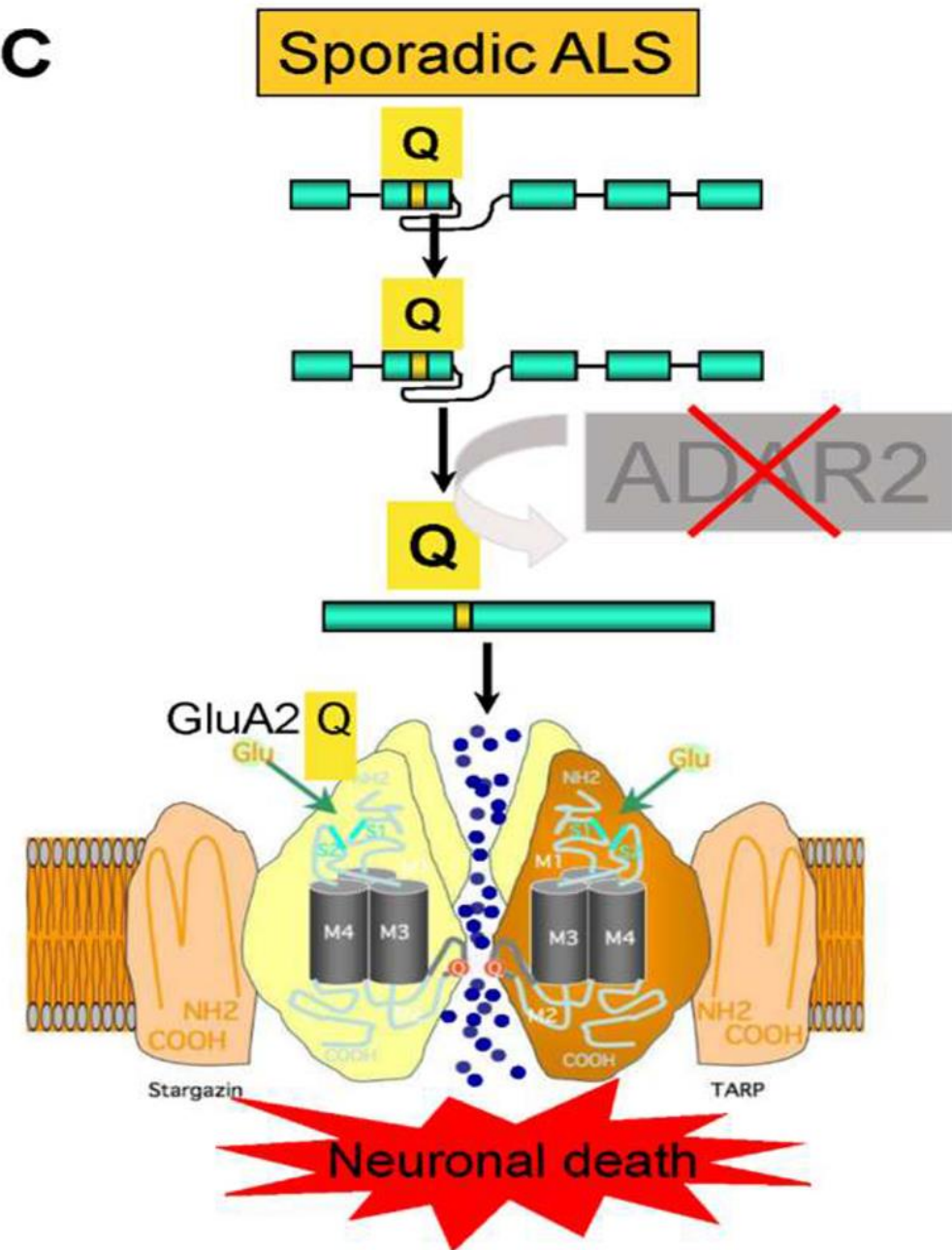


mSOD1 afeta a função da cadeia transportadora de elétrons, elevando a produção do ânion superóxido.

# Excitotoxicidade por Glutamato

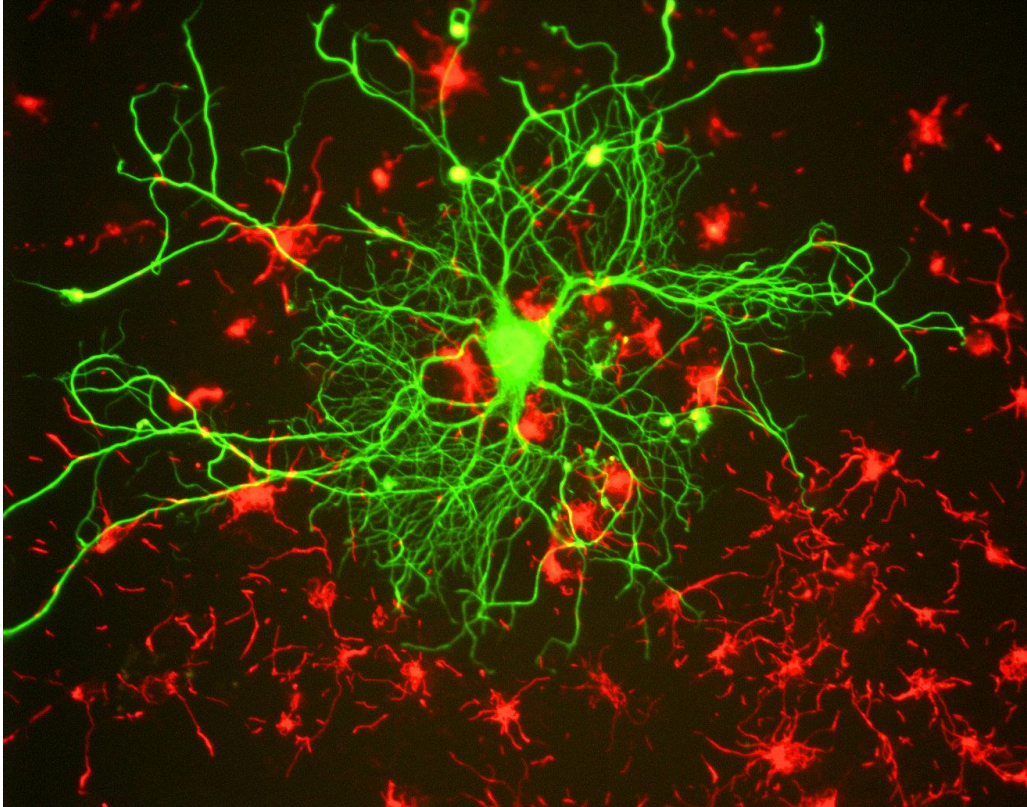


Van Den Bosch, L. et al, 2006.

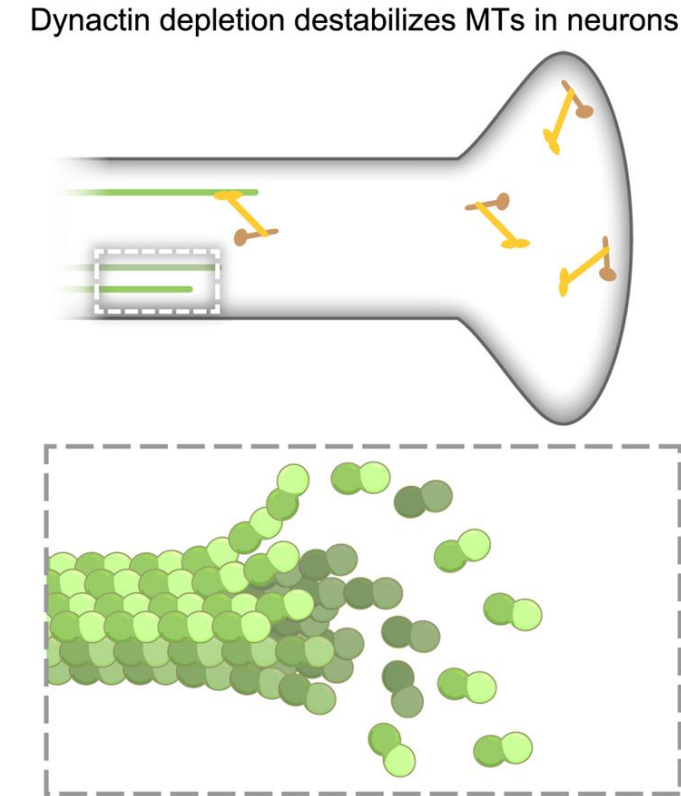
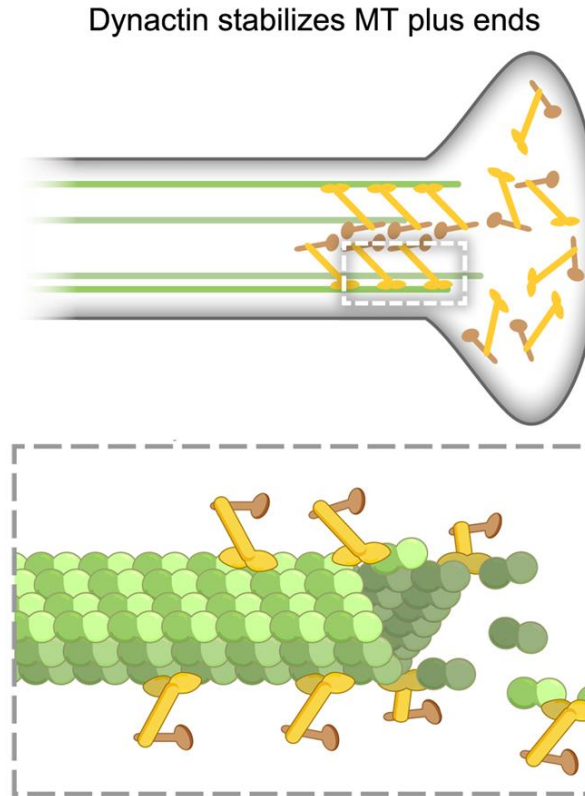
**B****C**



# Disfunção Axonal

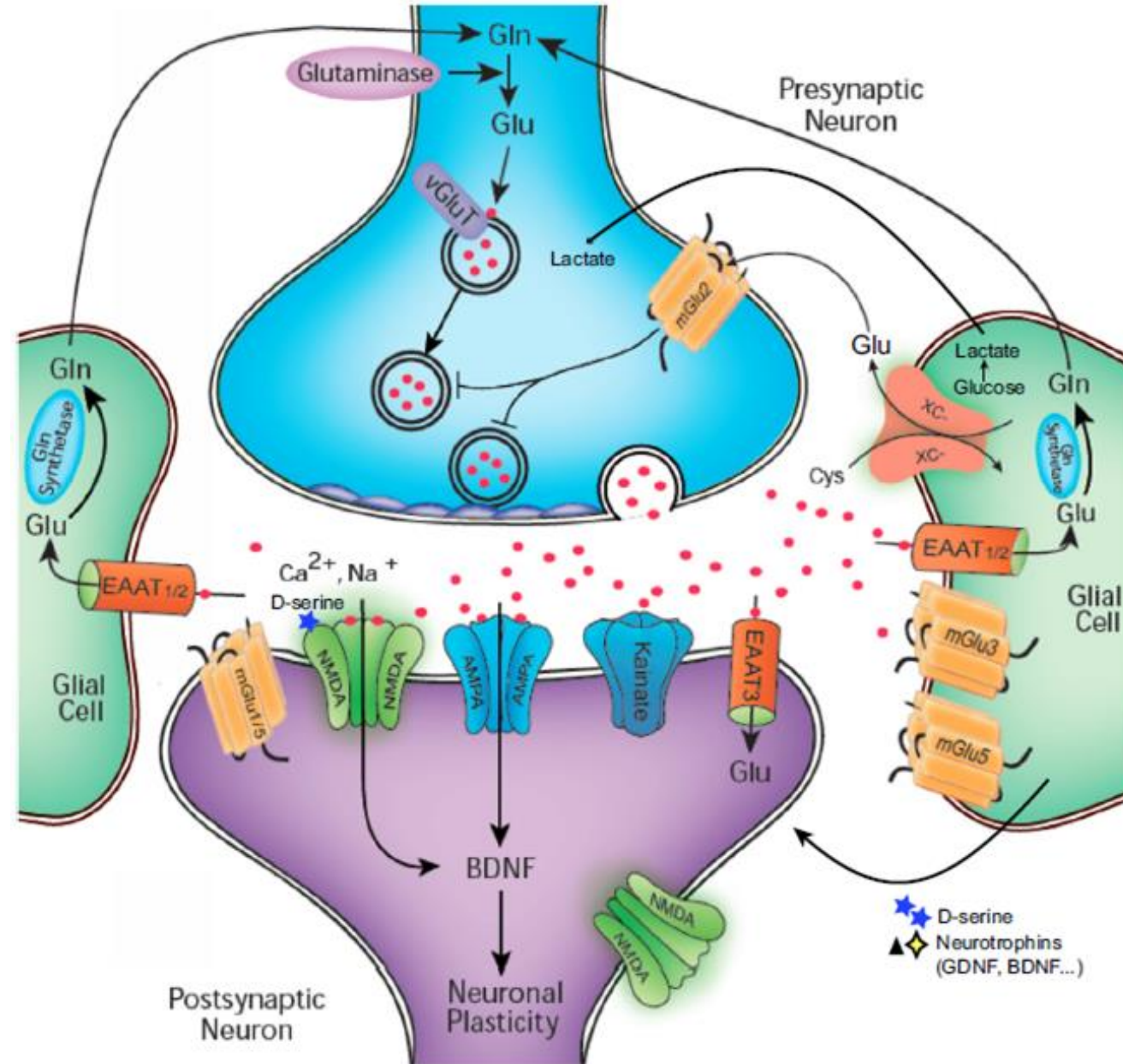


Rat brain cells grown in tissue culture and stained, in green, with an antibody to neurofilament subunit NF-L. -



Lazarus J.E., et al, 2013

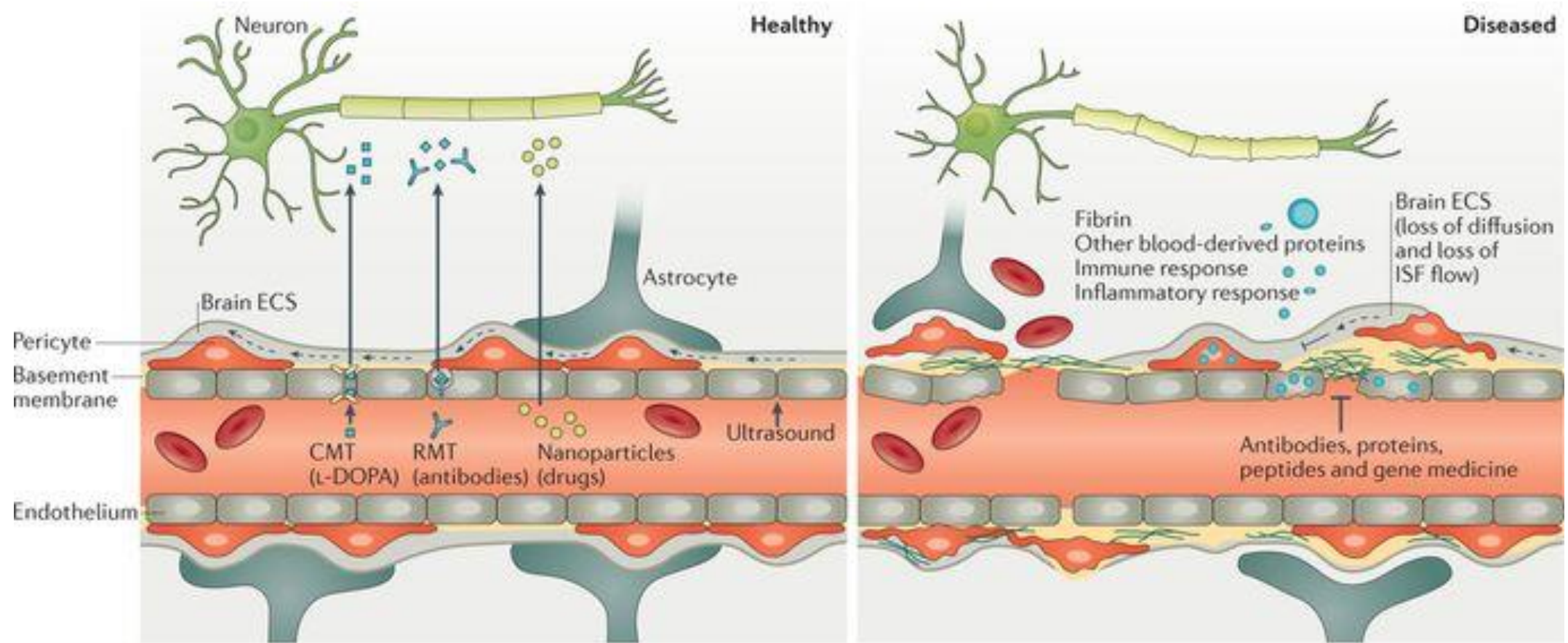
# Alteração da função dos Astrócitos



G. Sanacora and  
M. Banasr., et al, 2013



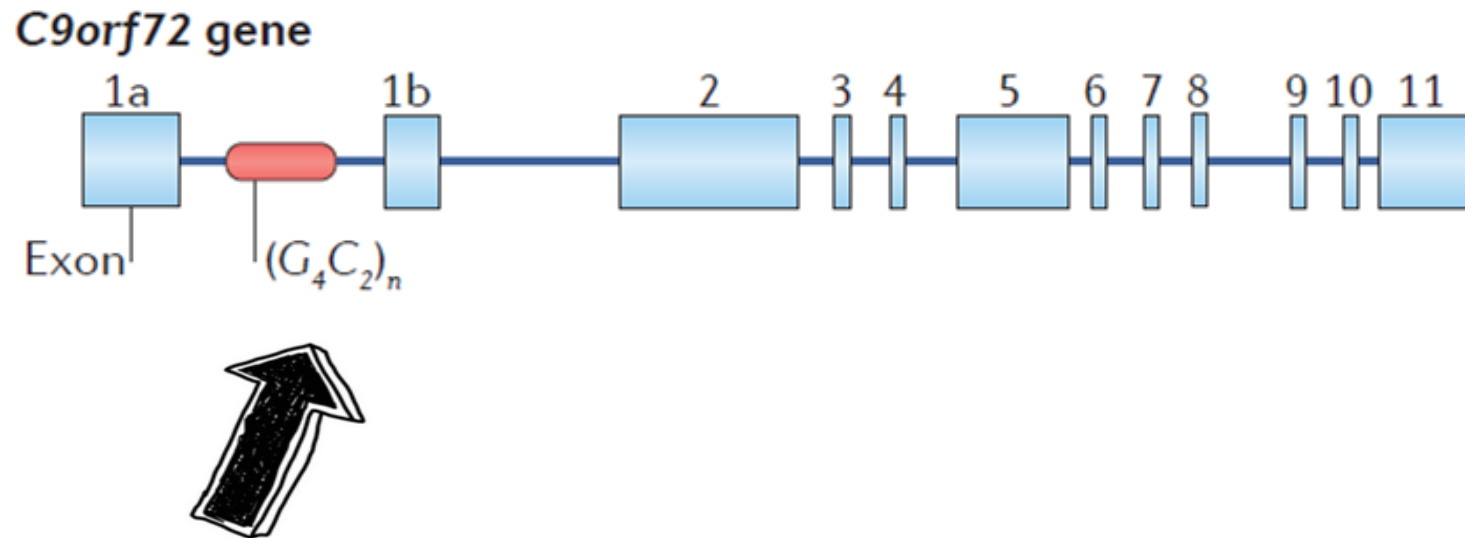
# Alterações neurovasculares



Nature Reviews | Neurology

Melanie D. Sweeney et al, 2018

# Transporte Núcleocitoplasmático?



Não repitam isso em casa

Apresentação disponível



SLIDES



# ESCLEROSE LATERAL AMIOTRÓFICA



VÍDEO

# Referências

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