

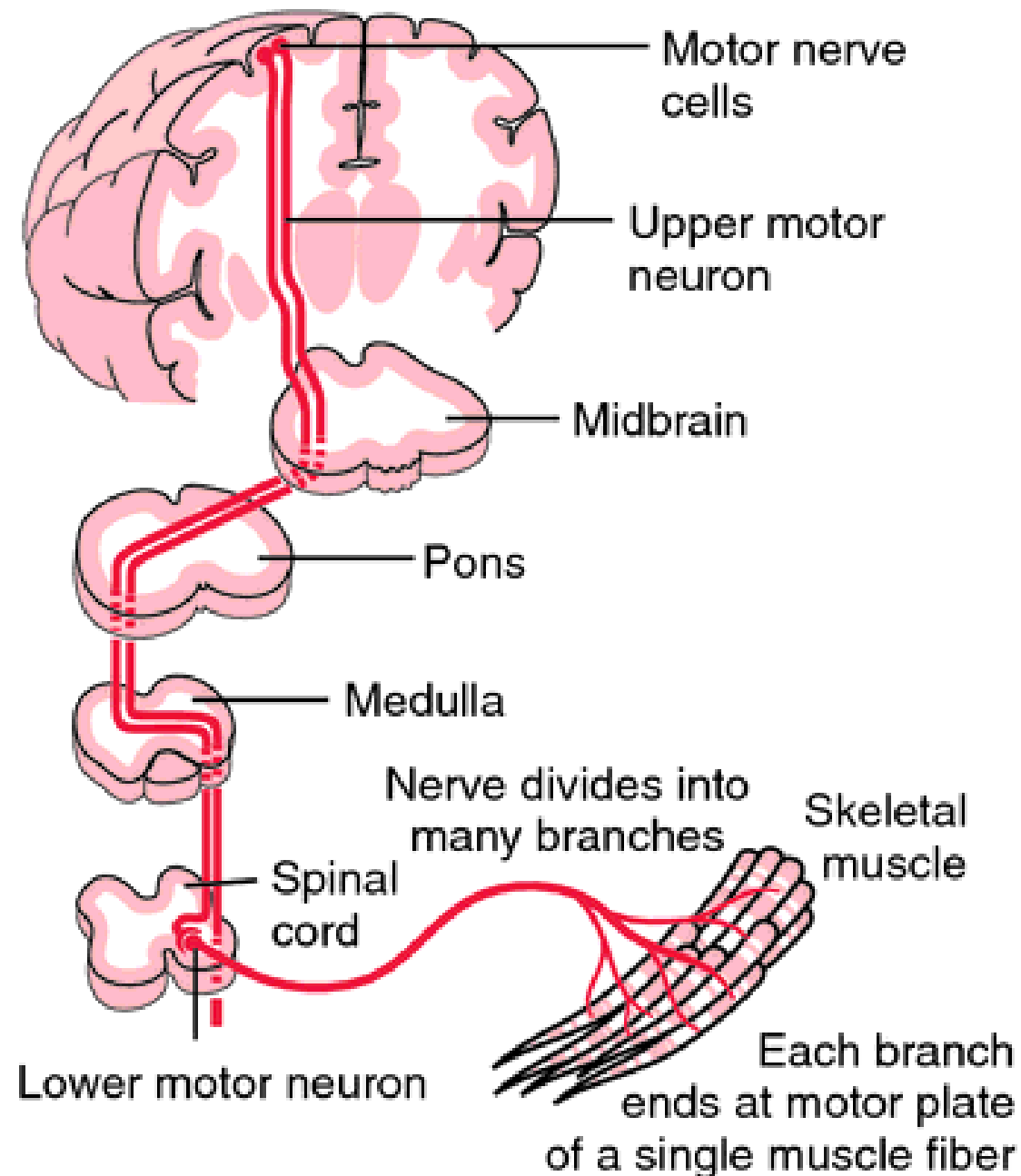


ESCLEROSE LATERAL AMIOTRÓFICA

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

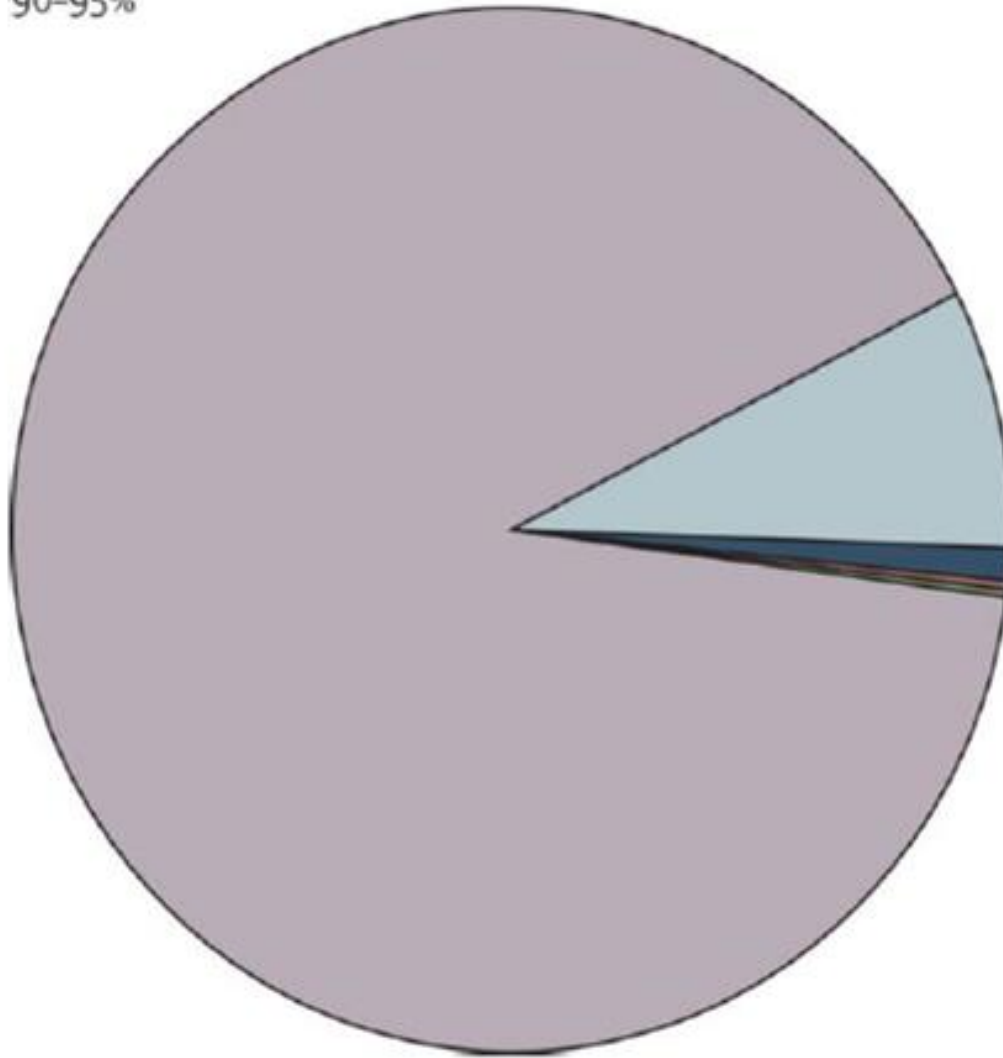


ESPORÁDICA

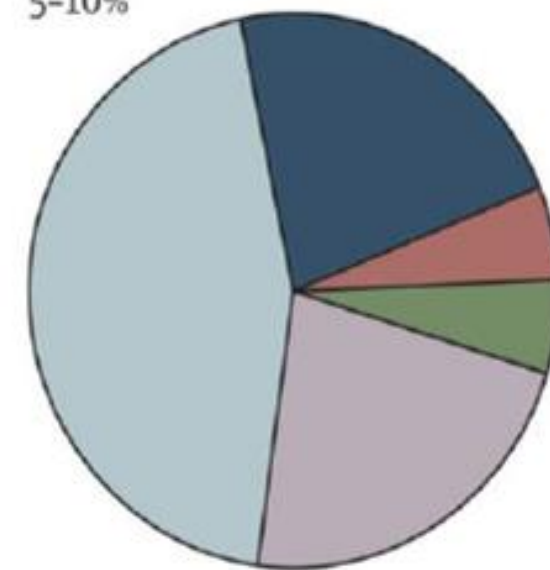


FAMILIAR

Apparently sporadic ALS
90-95%

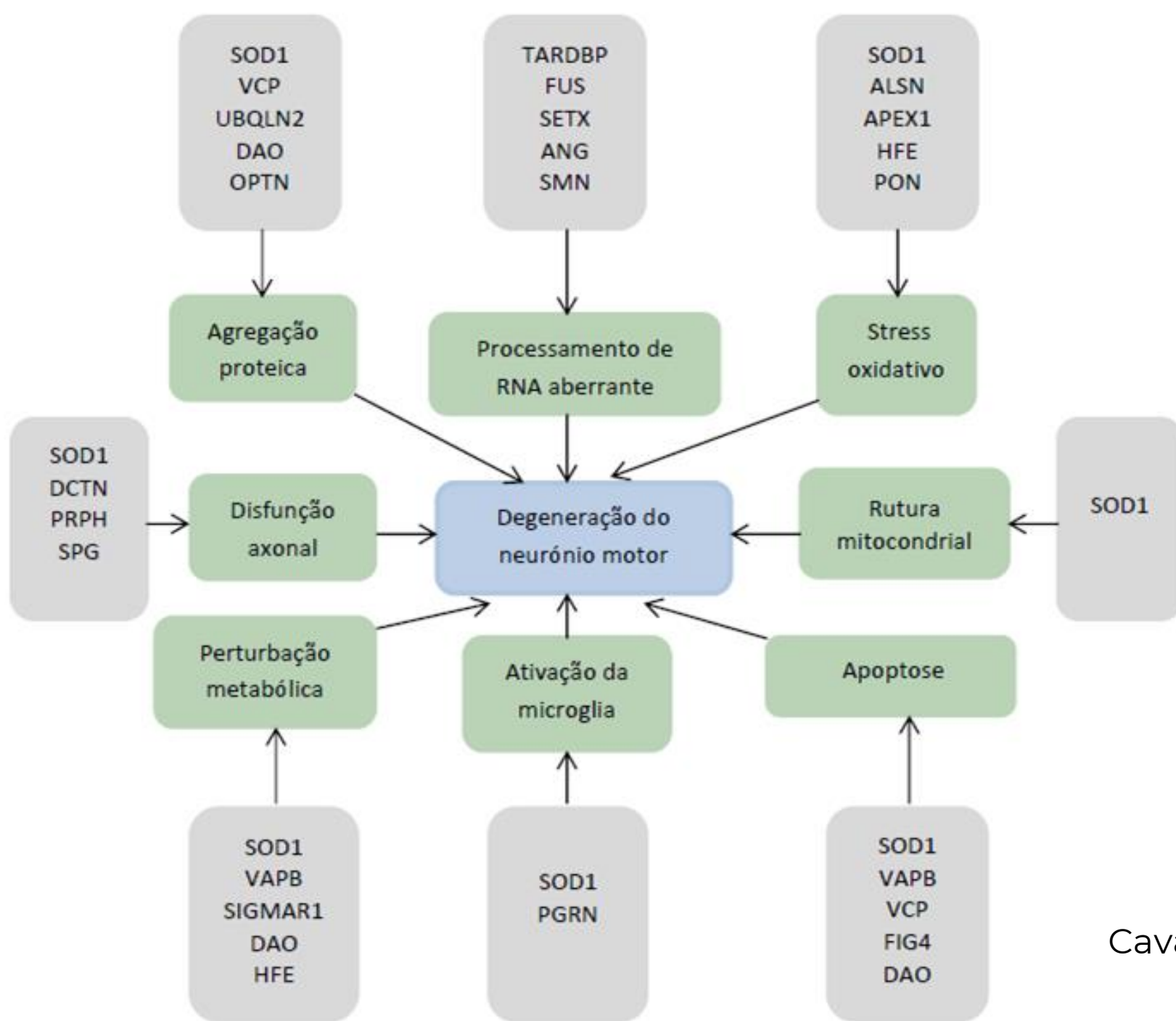


Familial ALS
5-10%



- SOD1
- TDP-43
- FUS
- Unknown
- C9orf72

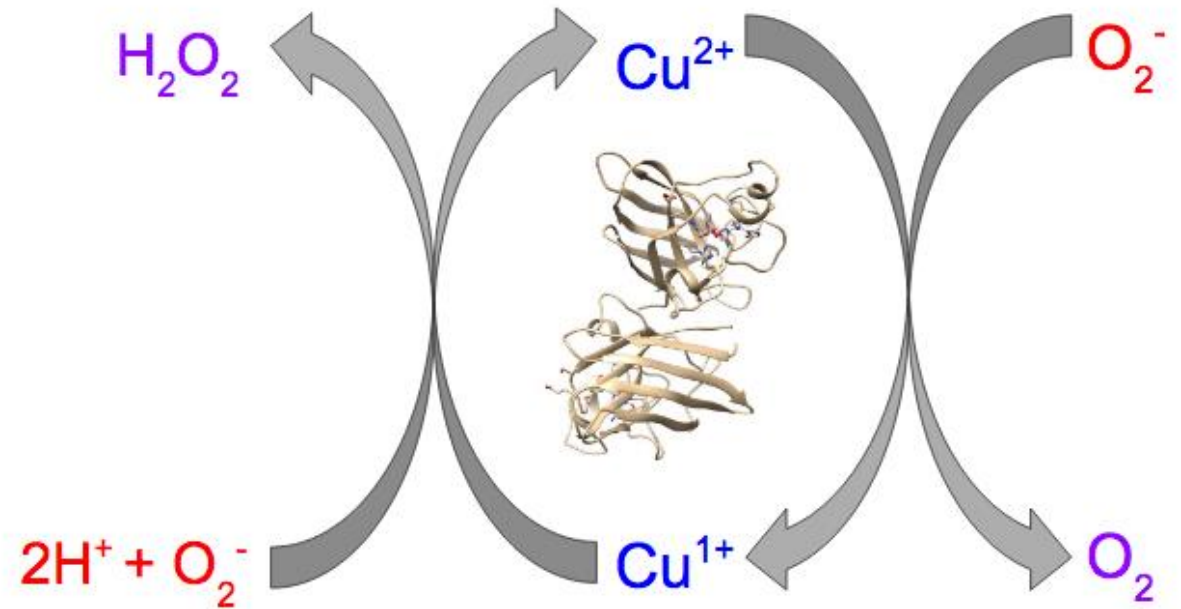
FISIOPATOLOGIA



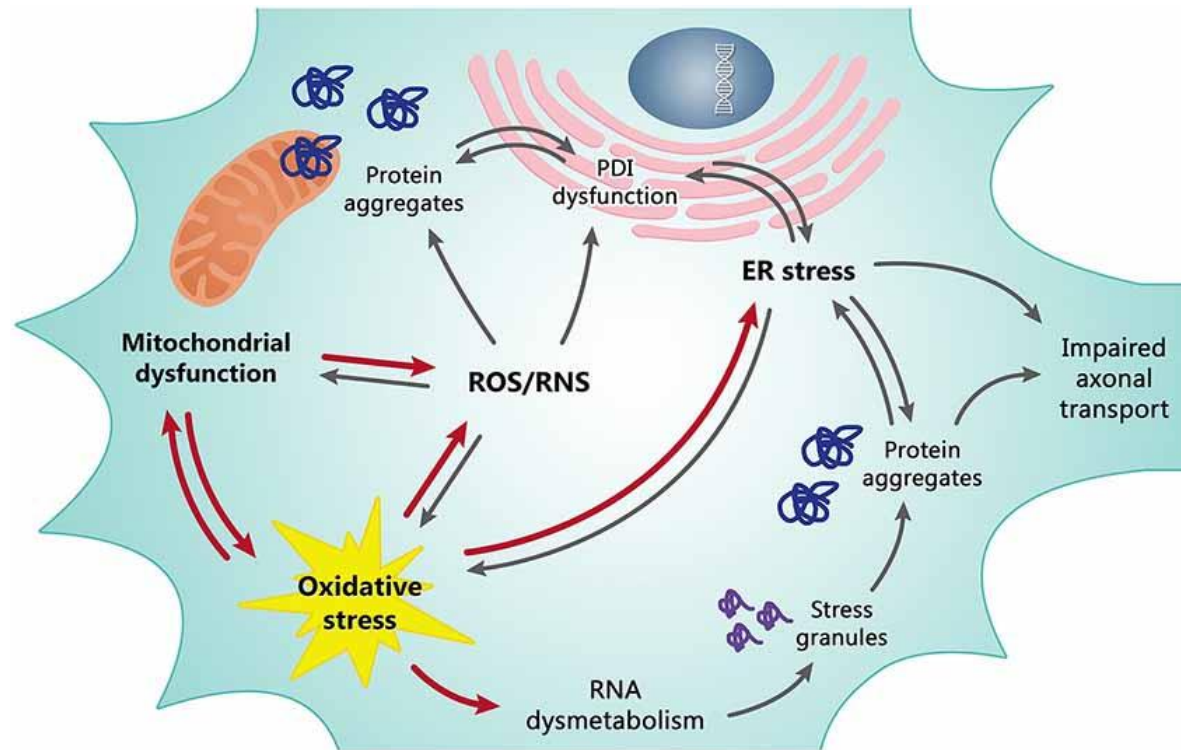
Stress Oxidativo

SOD1

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

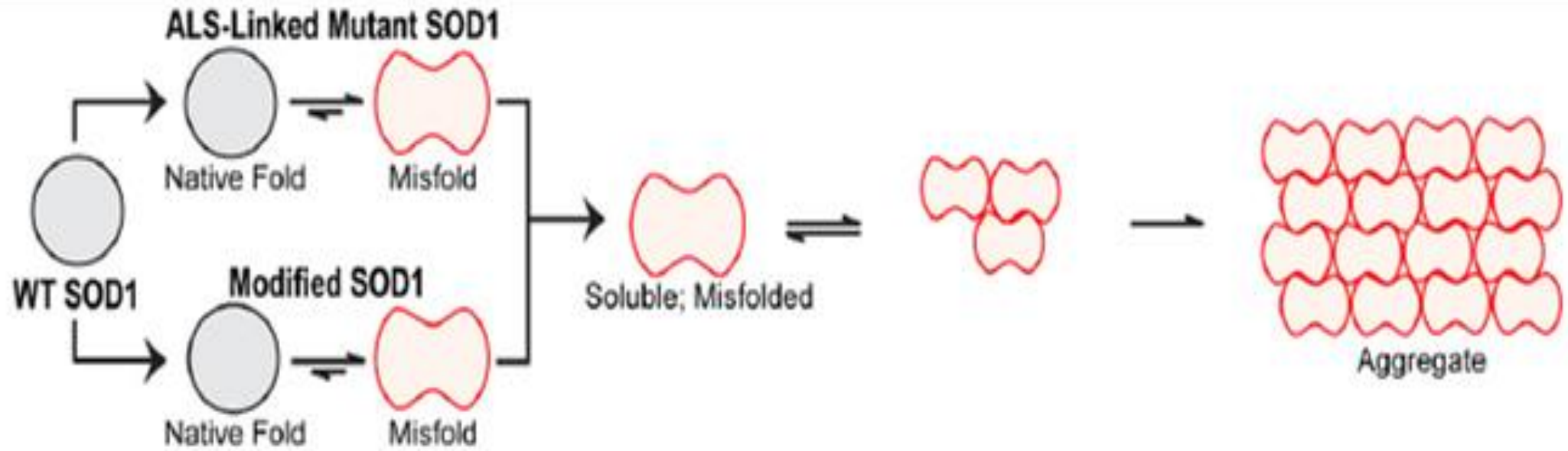


Stress Oxidativo



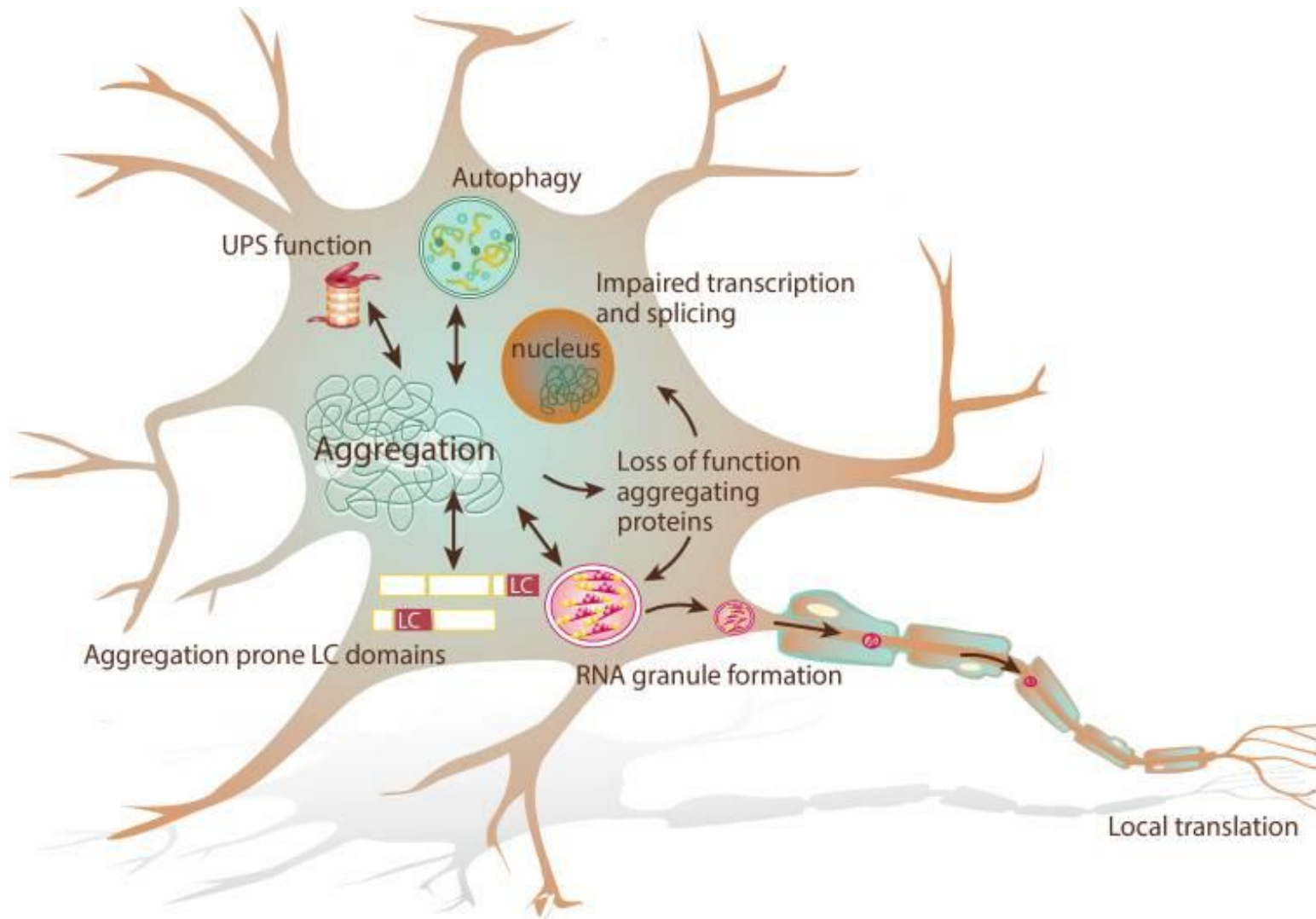
Carri et al, 2015

Agregação Proteica



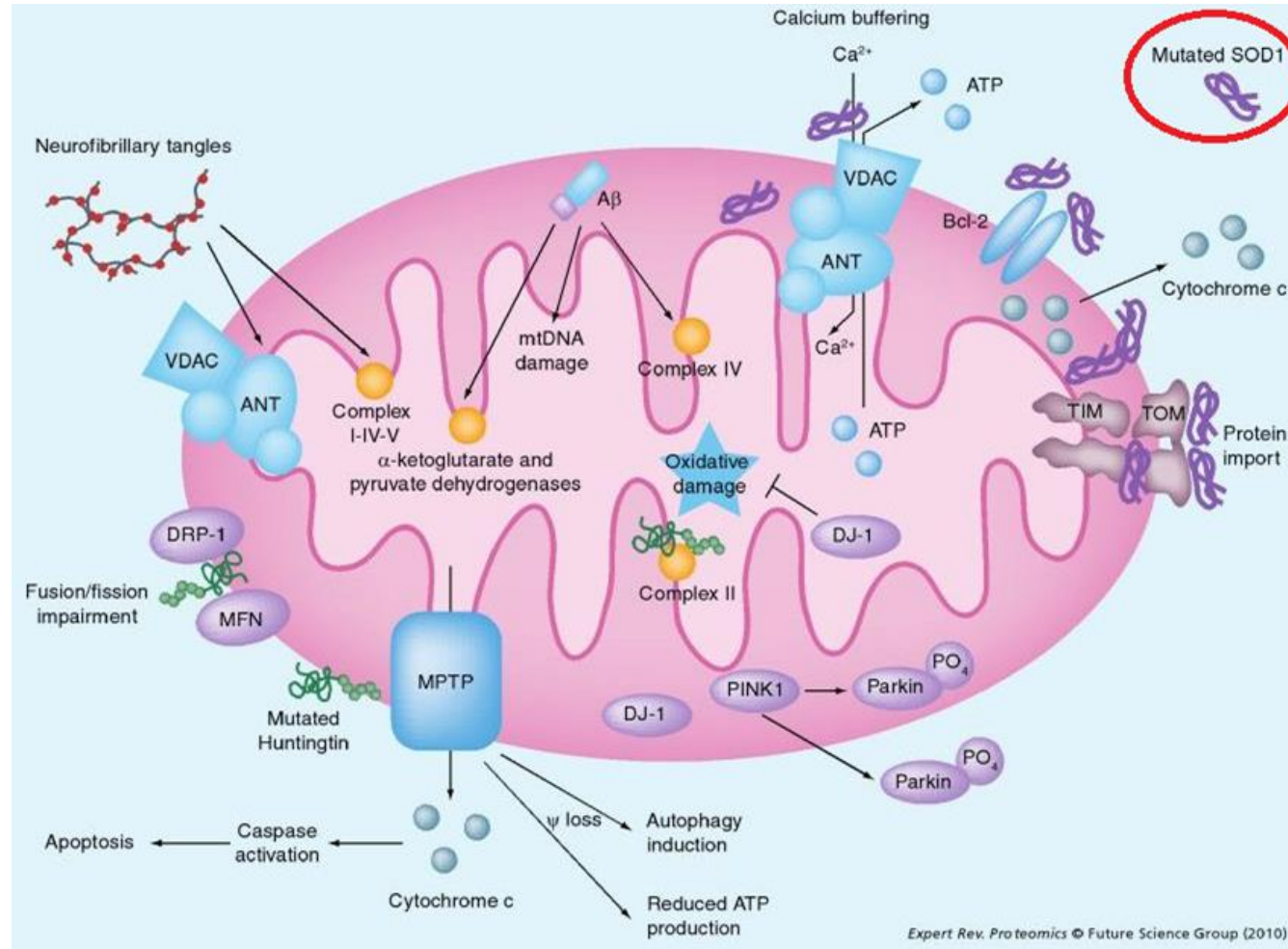
Rotunno and Bosco , 2013

Agregação Proteica



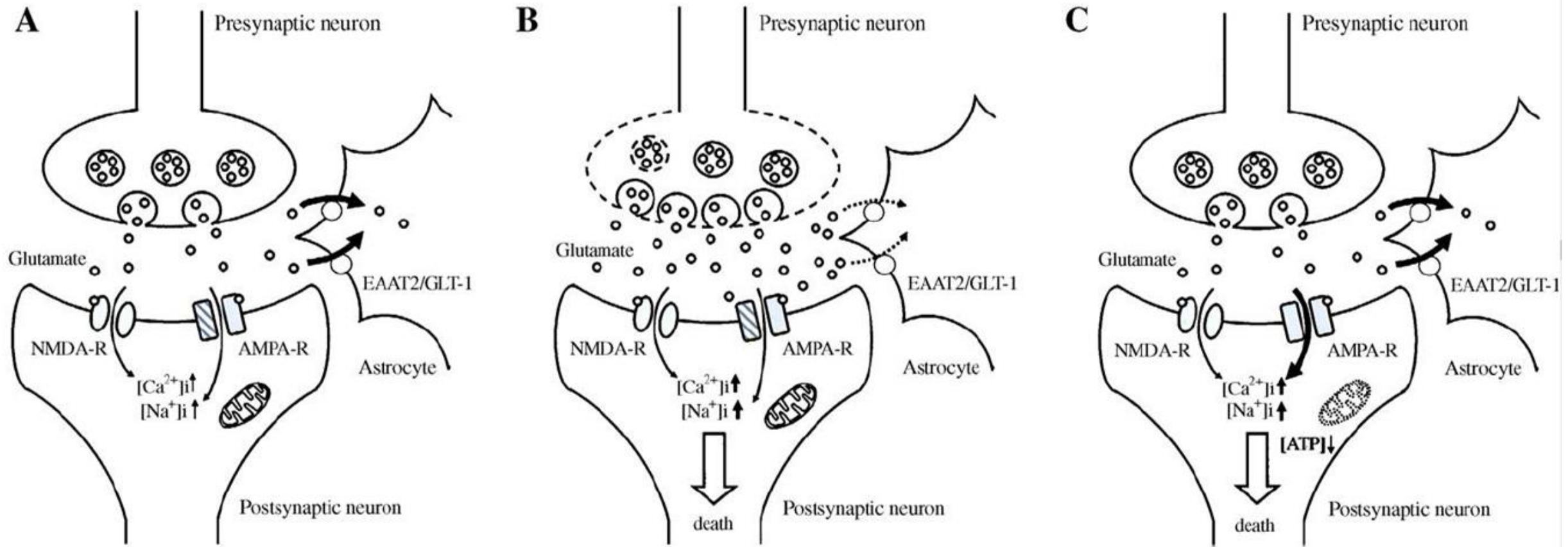
Blokhuis et al, 2013

Disfunção Mitochondrial

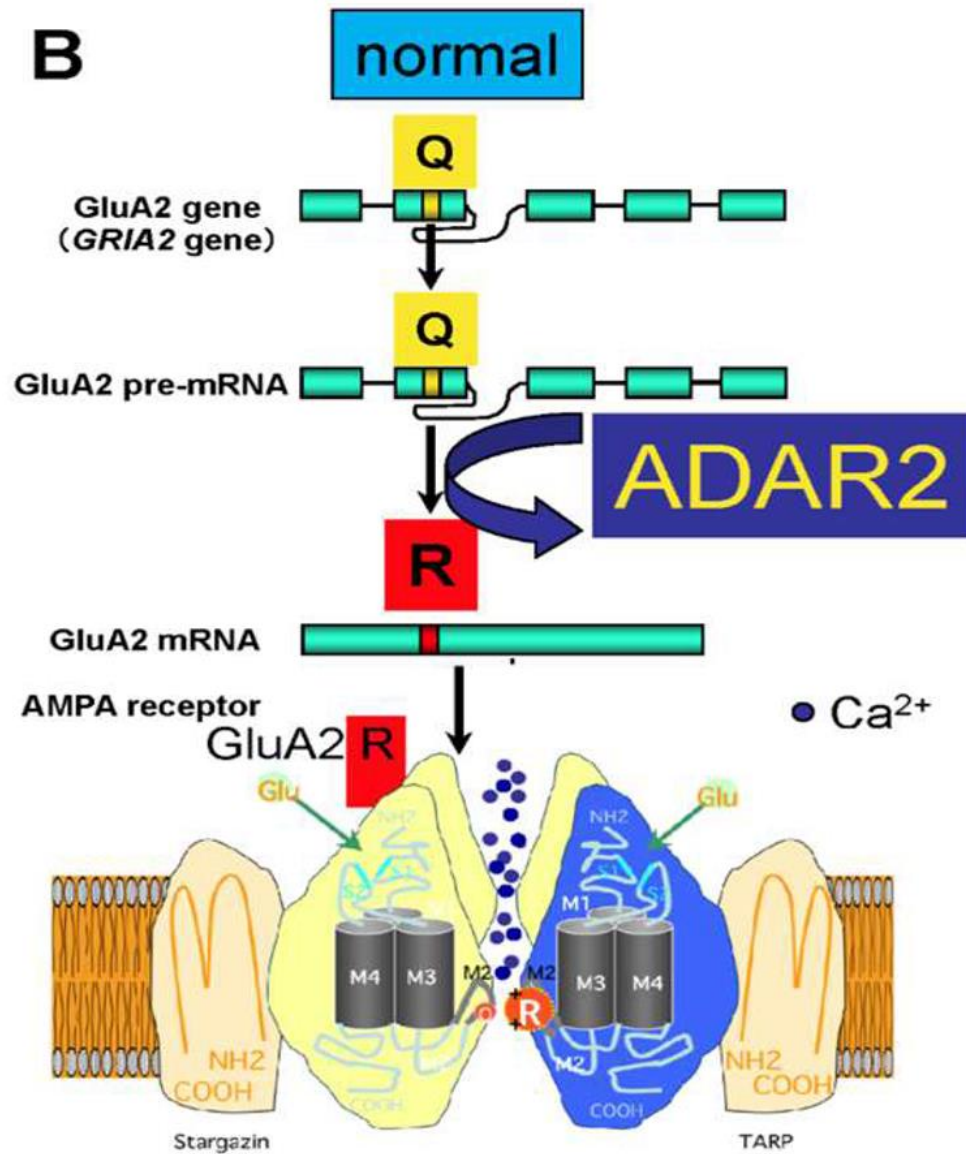
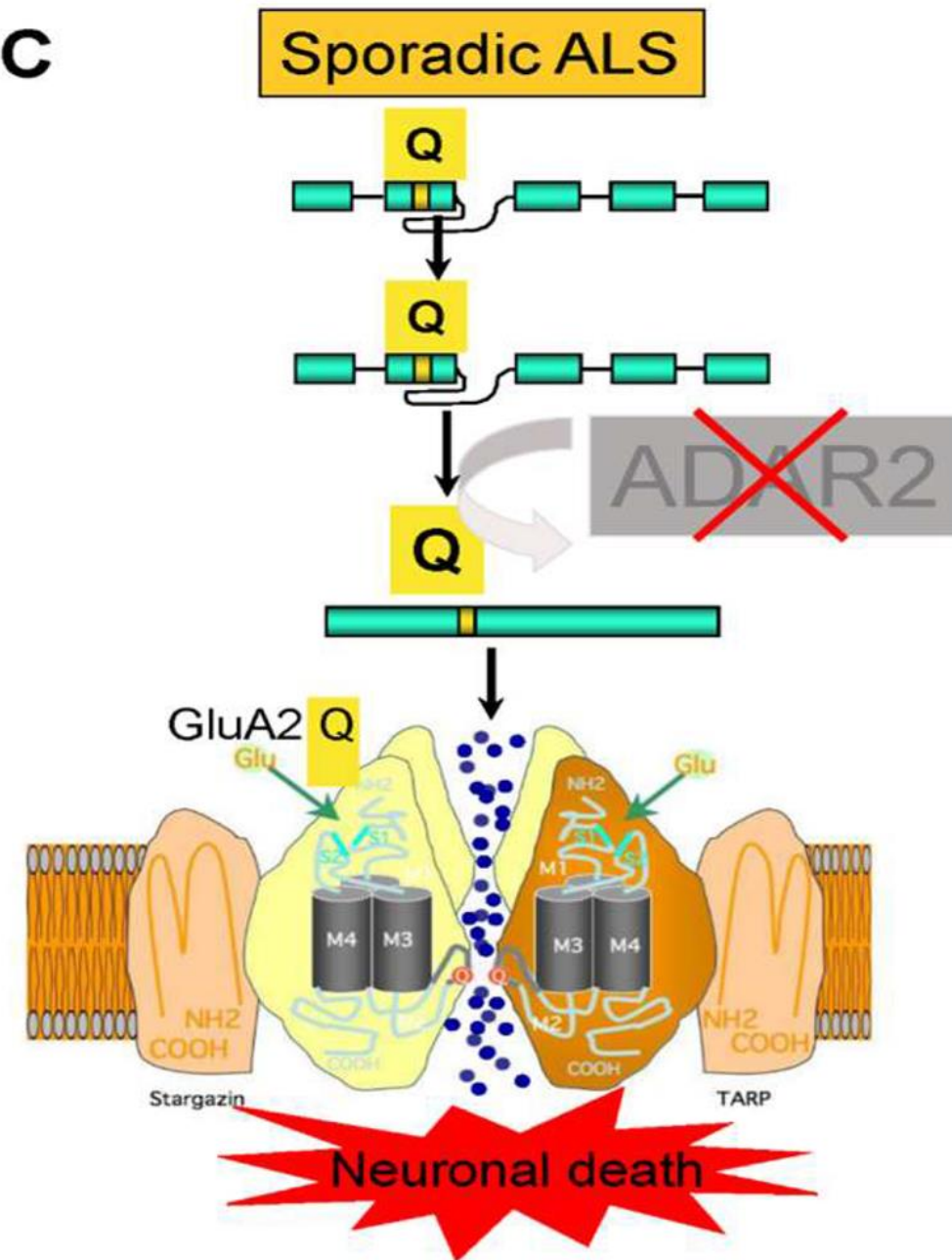


mSOD1 afeta a função da cadeia transportadora de elétrons, elevando a produção do ânion superóxido.

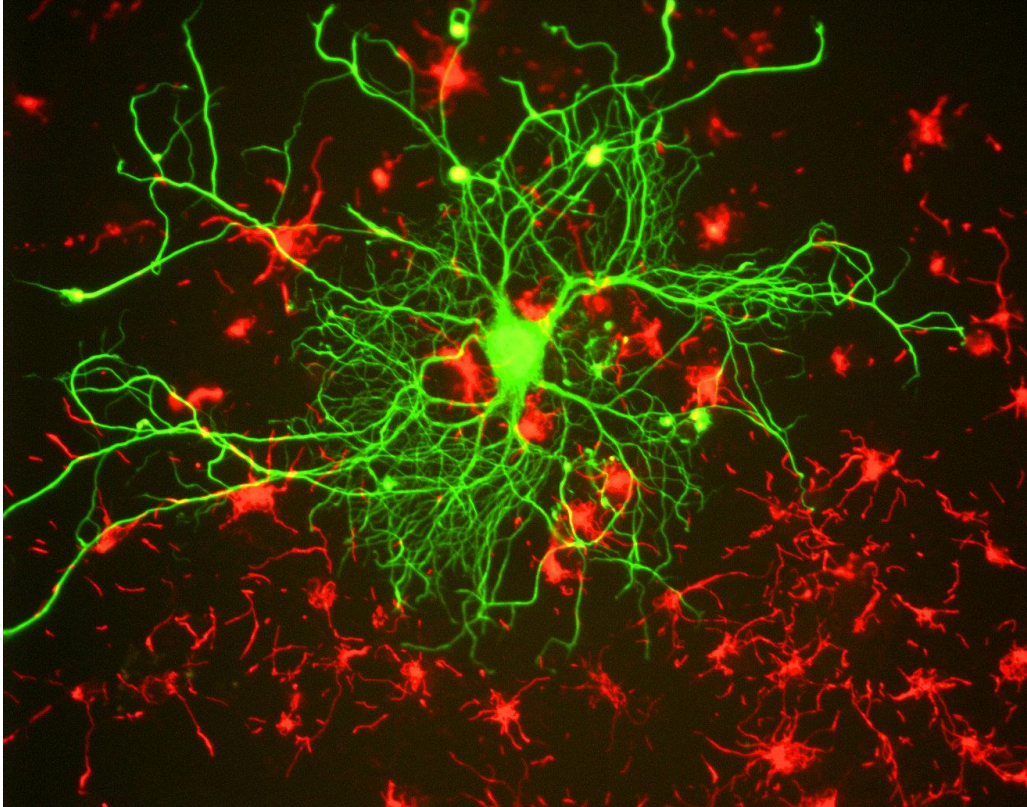
Excitotoxicidade por Glutamato



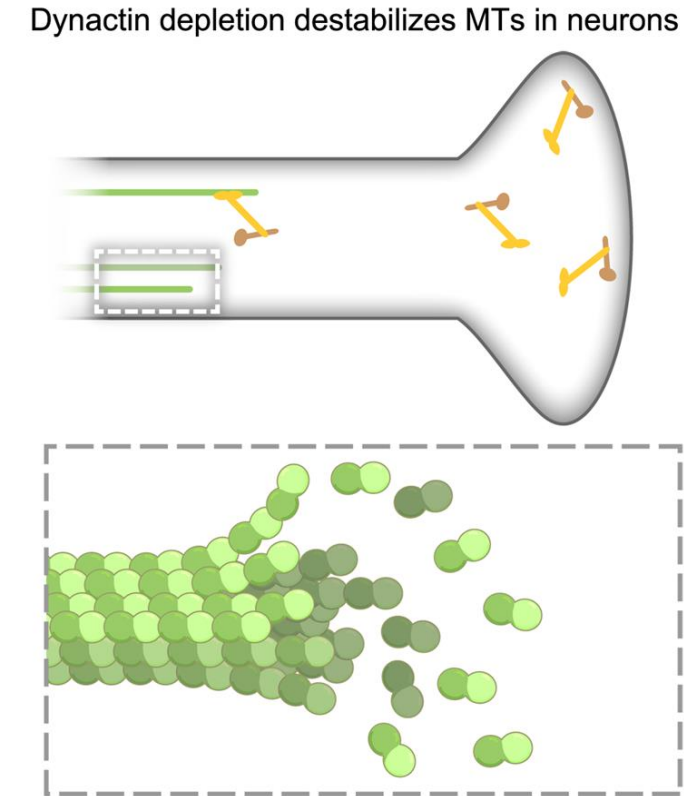
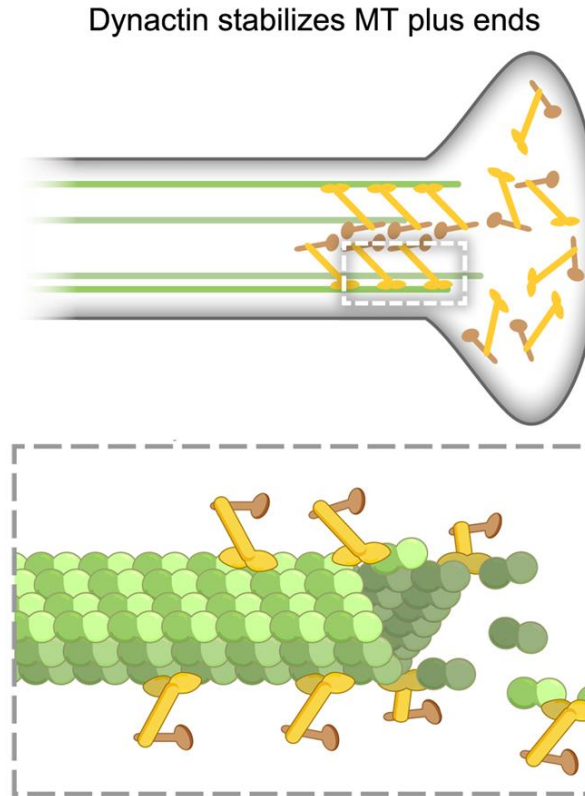
Van Den Bosch, L. et al, 2006.

B**C**

Disfunção Axonal

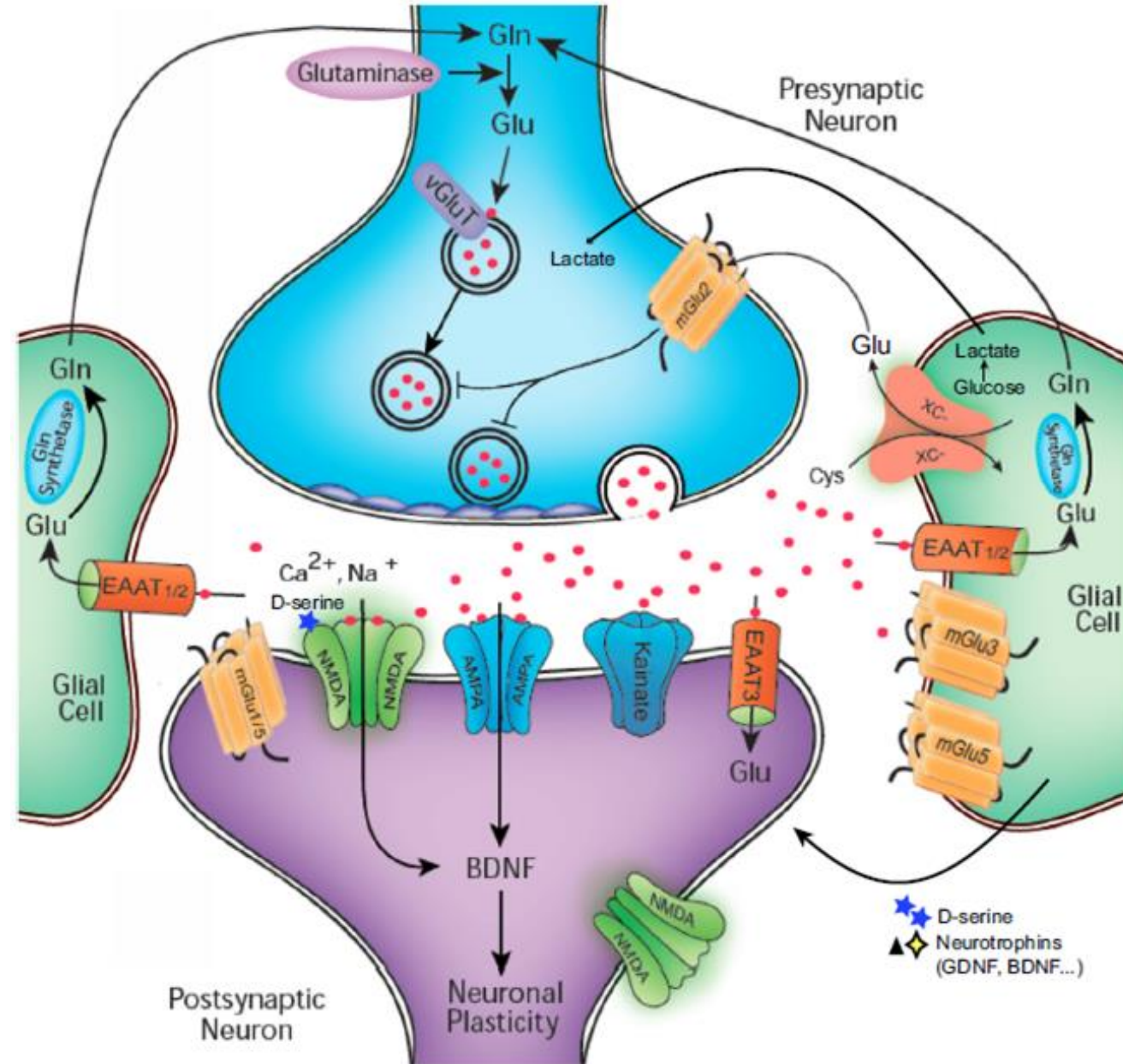


Rat brain cells grown in tissue culture and stained, in green, with an antibody to neurofilament subunit NF-L. -



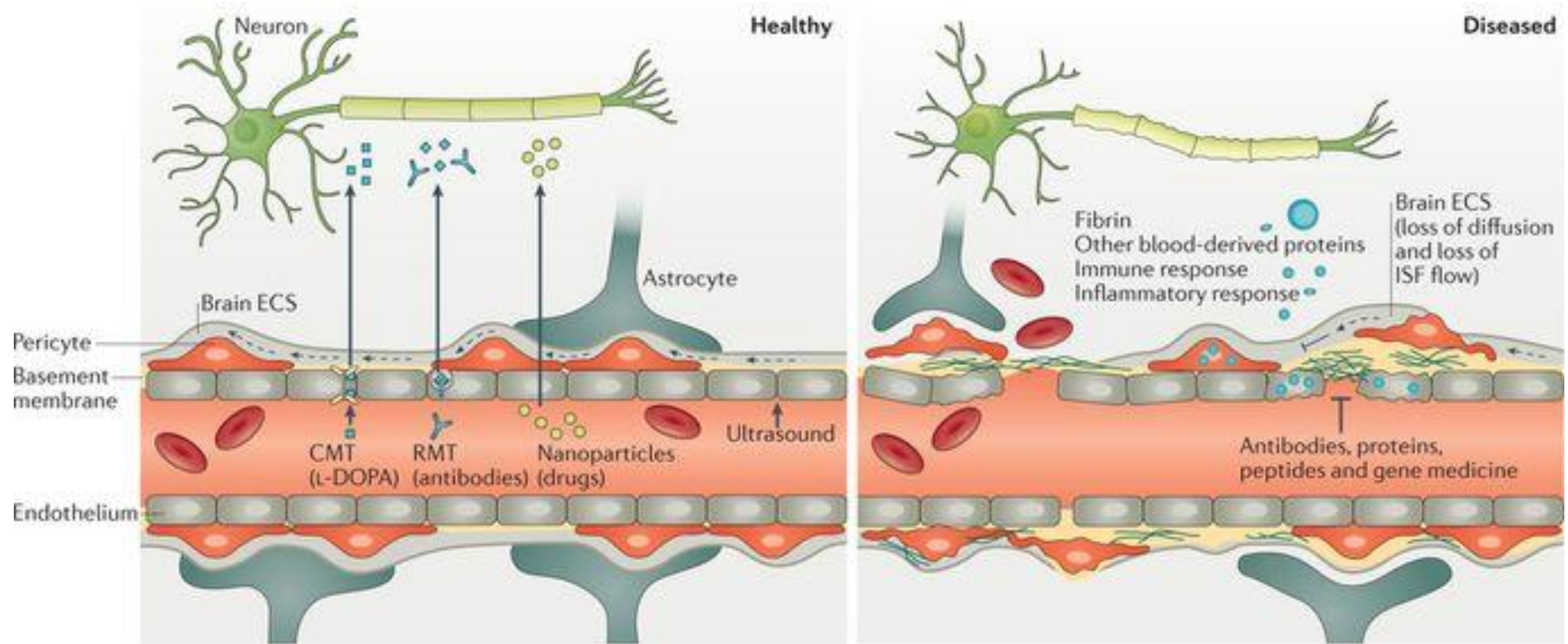
Lazarus J.E., et al, 2013

Alteração da função dos Astrócitos



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

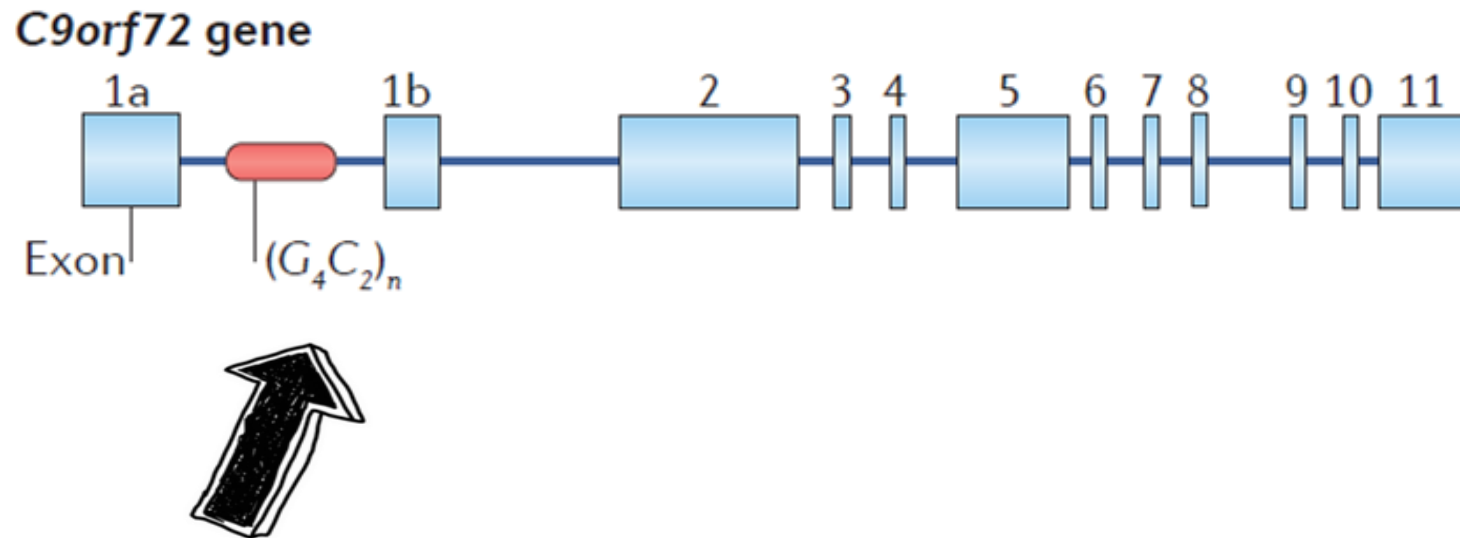
Alterações neurovasculares



Nature Reviews | Neurology

Melanie D. Sweeney et al, 2018

Transporte Núcleocitoplasmático?



Não repitam isso em casa

Apresentação disponível



SLIDES



ESCLEROSE LATERAL AMIOTRÓFICA



VÍDEO

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