

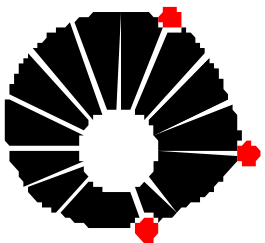
Ataxia de Friedreich

Prof. Marcondes França

Departamento de Neurologia

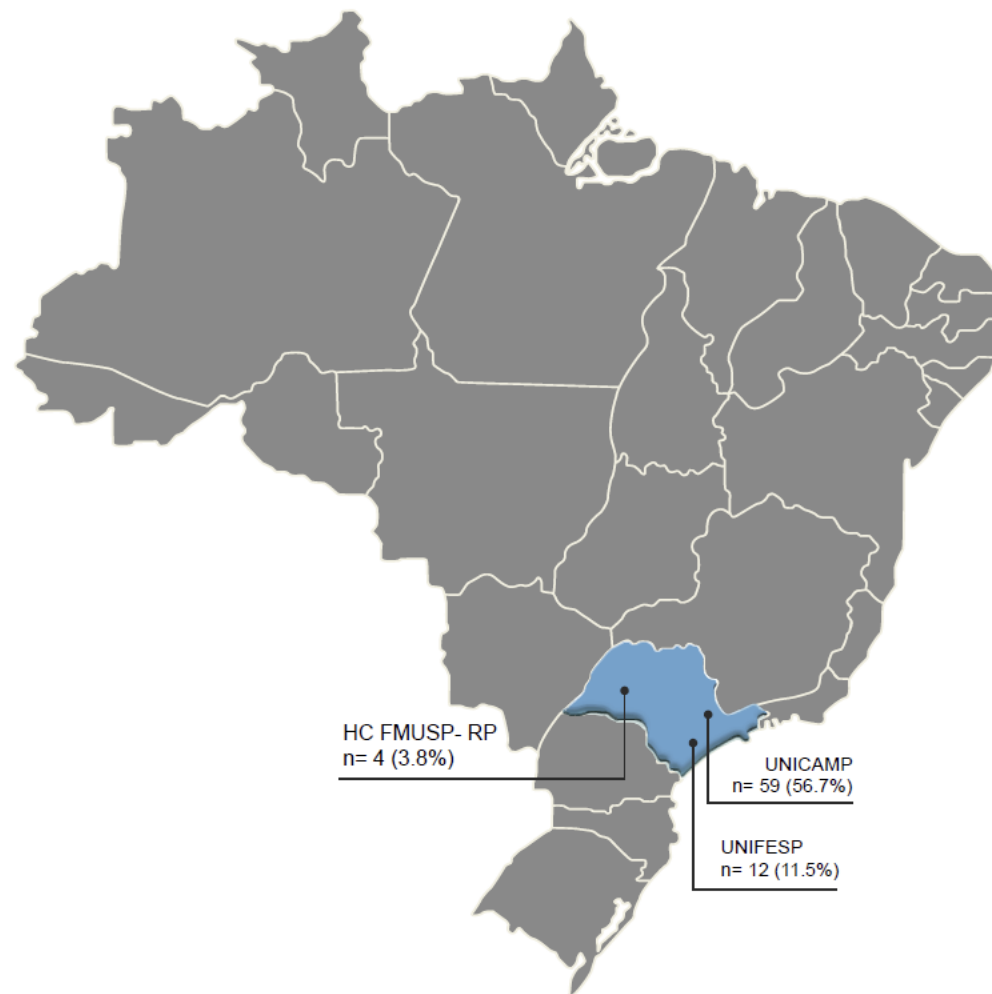
Universidade de Campinas (UNICAMP)

Brasil



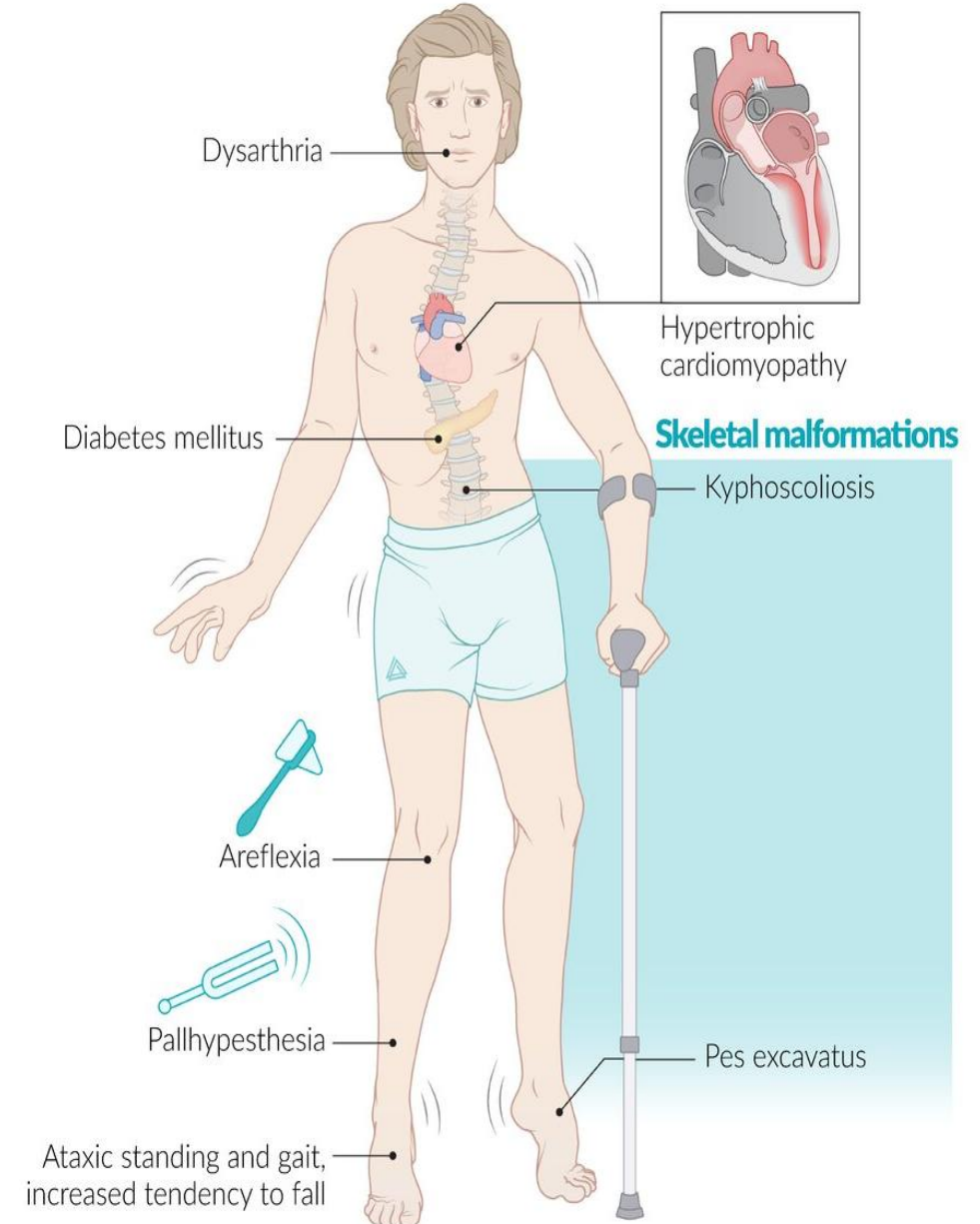
Ataxia de Friedreich: Prevalência no Brasil

- Levantamento no estado de São Paulo
- Múltiplas fontes de dado:
 - Centros de referência
 - Associação de ataxia (ABAHE)
 - Academia de Neurologia (ABN)
- **Prevalência = 0.36: 100,000**



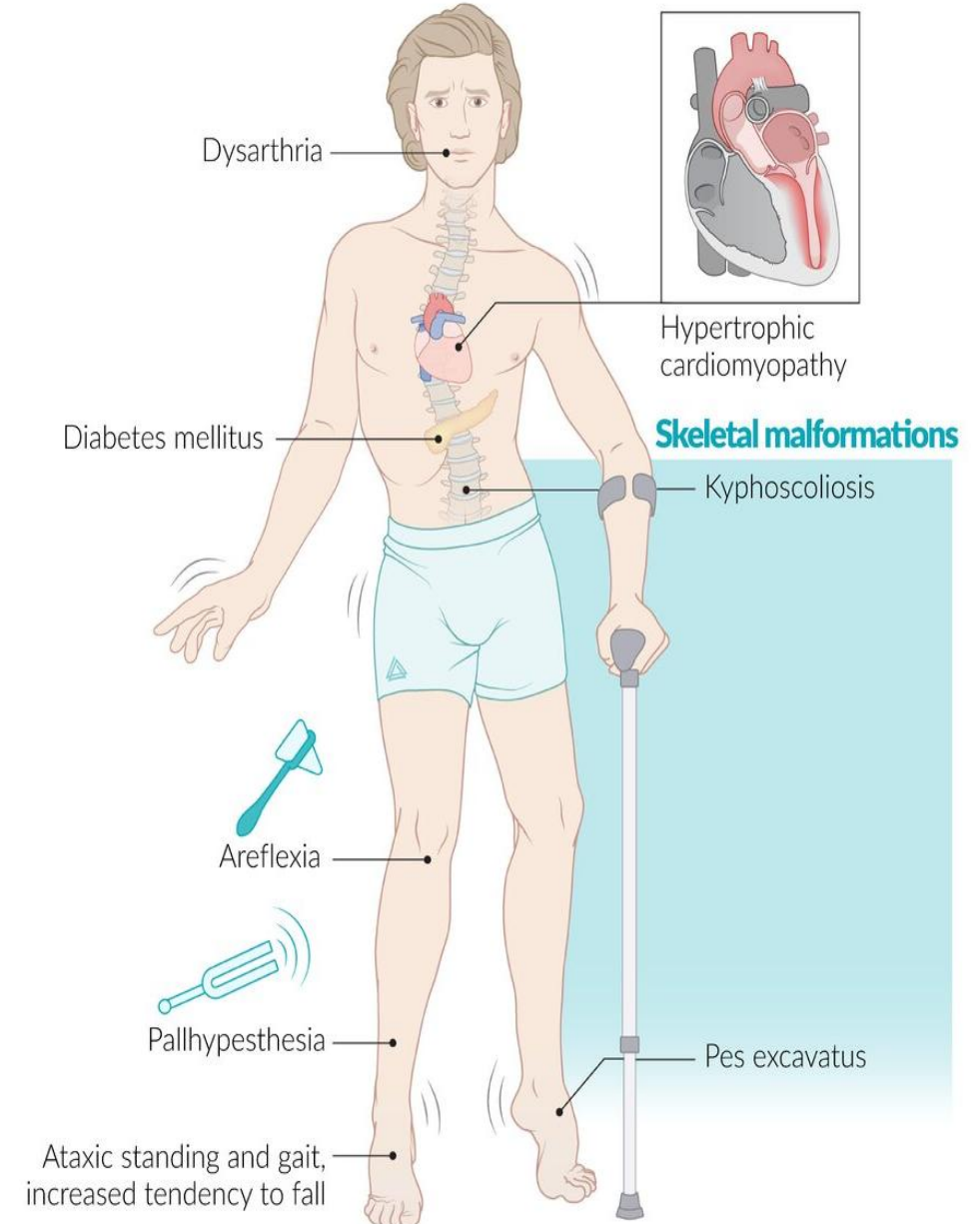
Ataxia de Friedreich: Características-chave

- Base genética
- Doença neurodegenerativa
- Envolvimento sistêmico

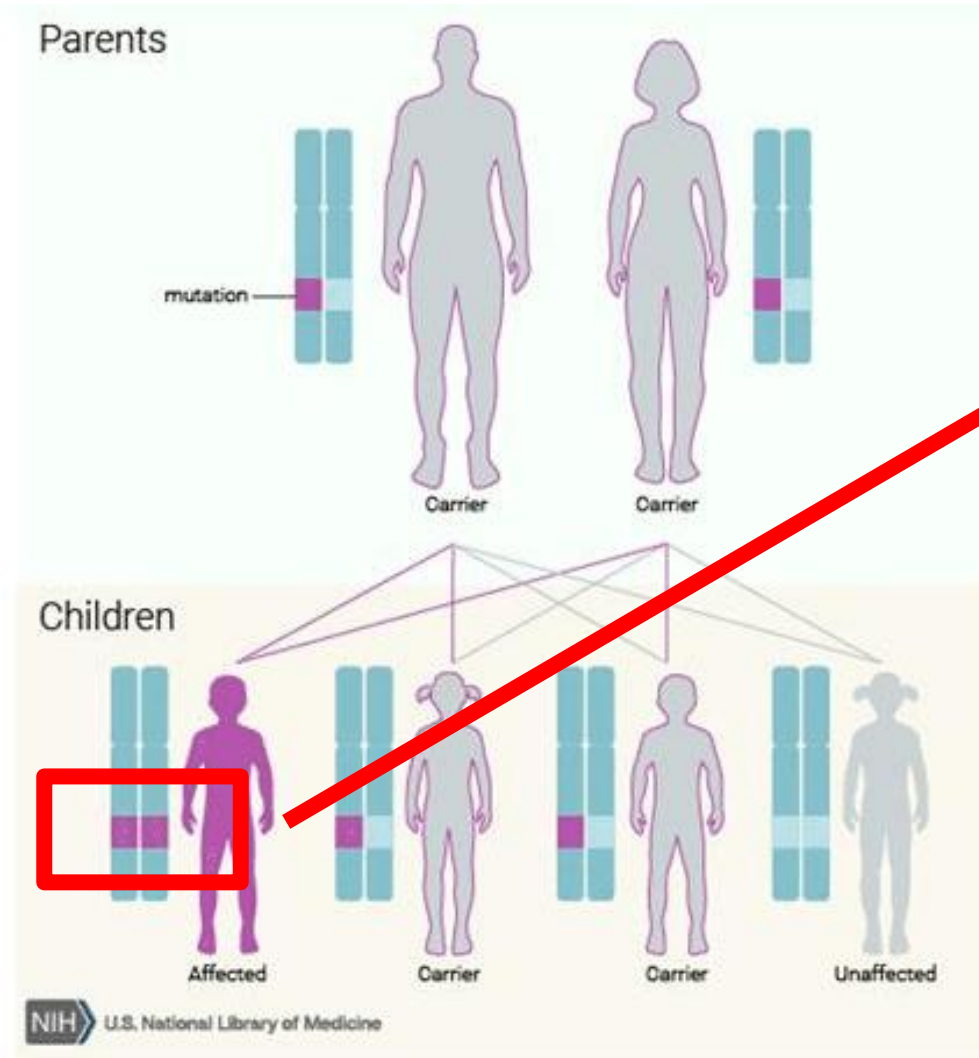


Ataxia de Friedreich: Características-chave

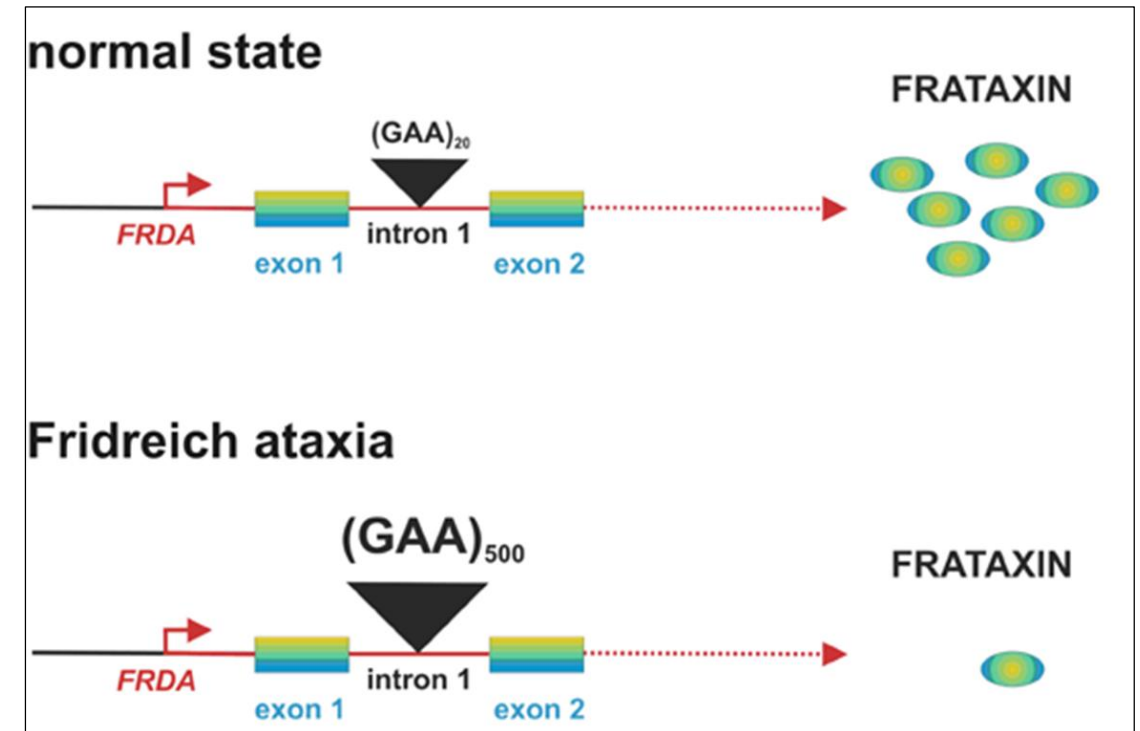
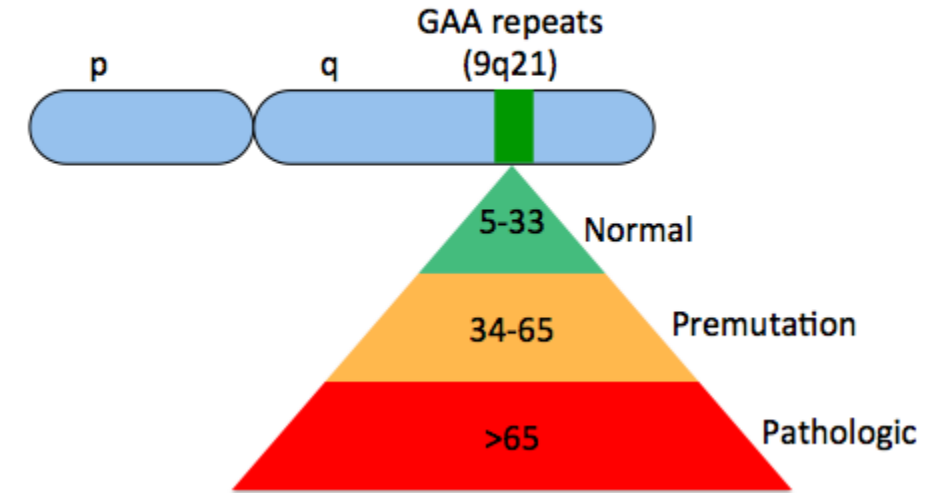
- Base genética
- Doença neurodegenerativa
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Ataxia de Friedreich: Genética

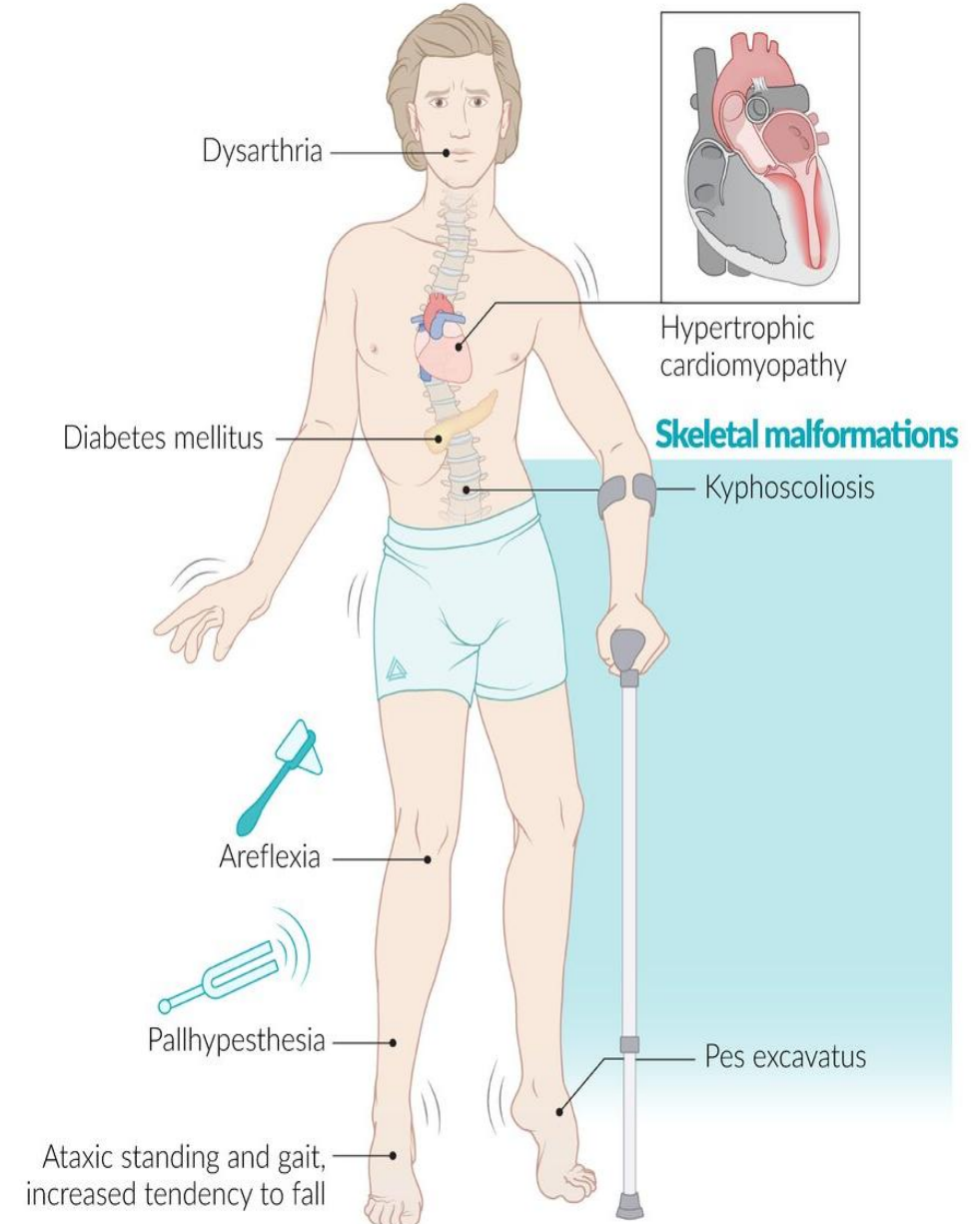


Friedreich ataxia

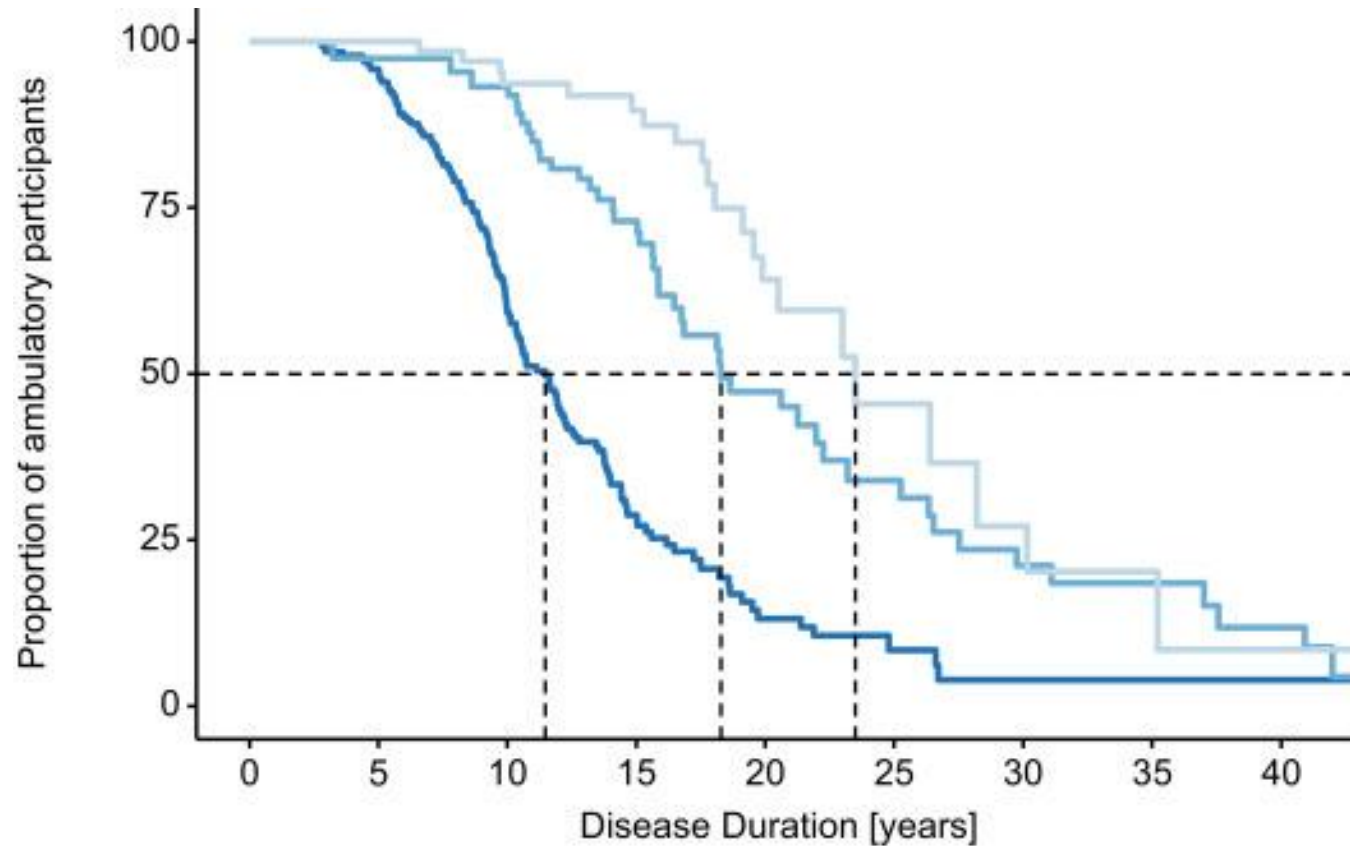


Ataxia de Friedreich: Características-chave

- Base genética
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Ataxia de Friedreich: Evolução



Perda definitiva de marcha:
Em média 10 anos após os
primeiros sintomas

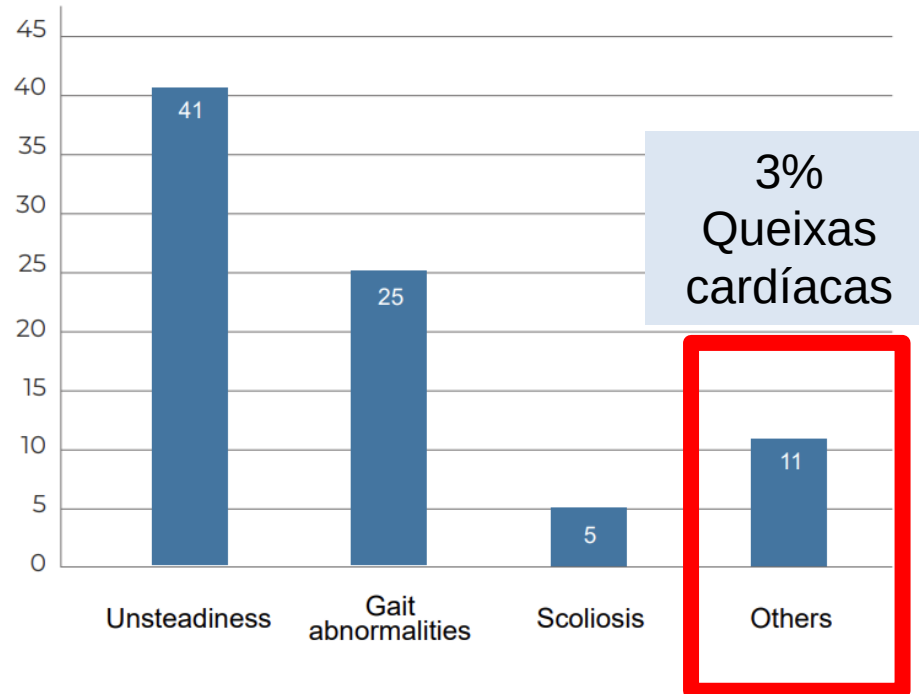
Number at risk										
Strata		Disease Duration [years]								
		0	5	10	15	20	25	30	35	40
	<15y	703	597	261	84	25	10	3	2	2
	15-24y	211	193	150	93	44	23	12	8	4
	>24y	107	102	82	66	31	13	6	3	1
		0	5	10	15	20	25	30	35	40

Ataxia de Friedreich: Jornada diagnóstica (Brasil)

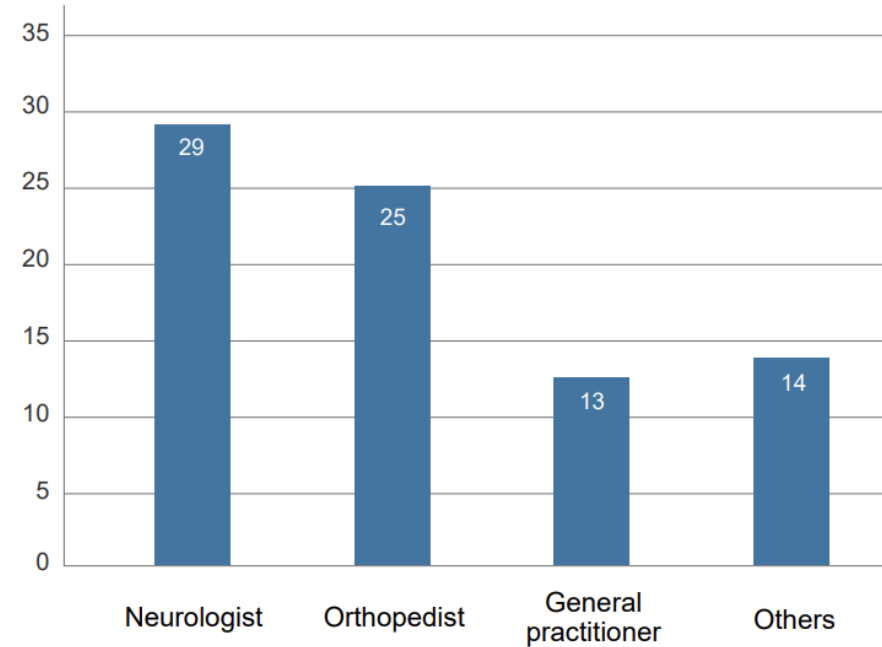
Atraso diagnóstico = 7.8 ± 6.7 anos

Erro diagnóstico = 46.3%

A- First symptom reported

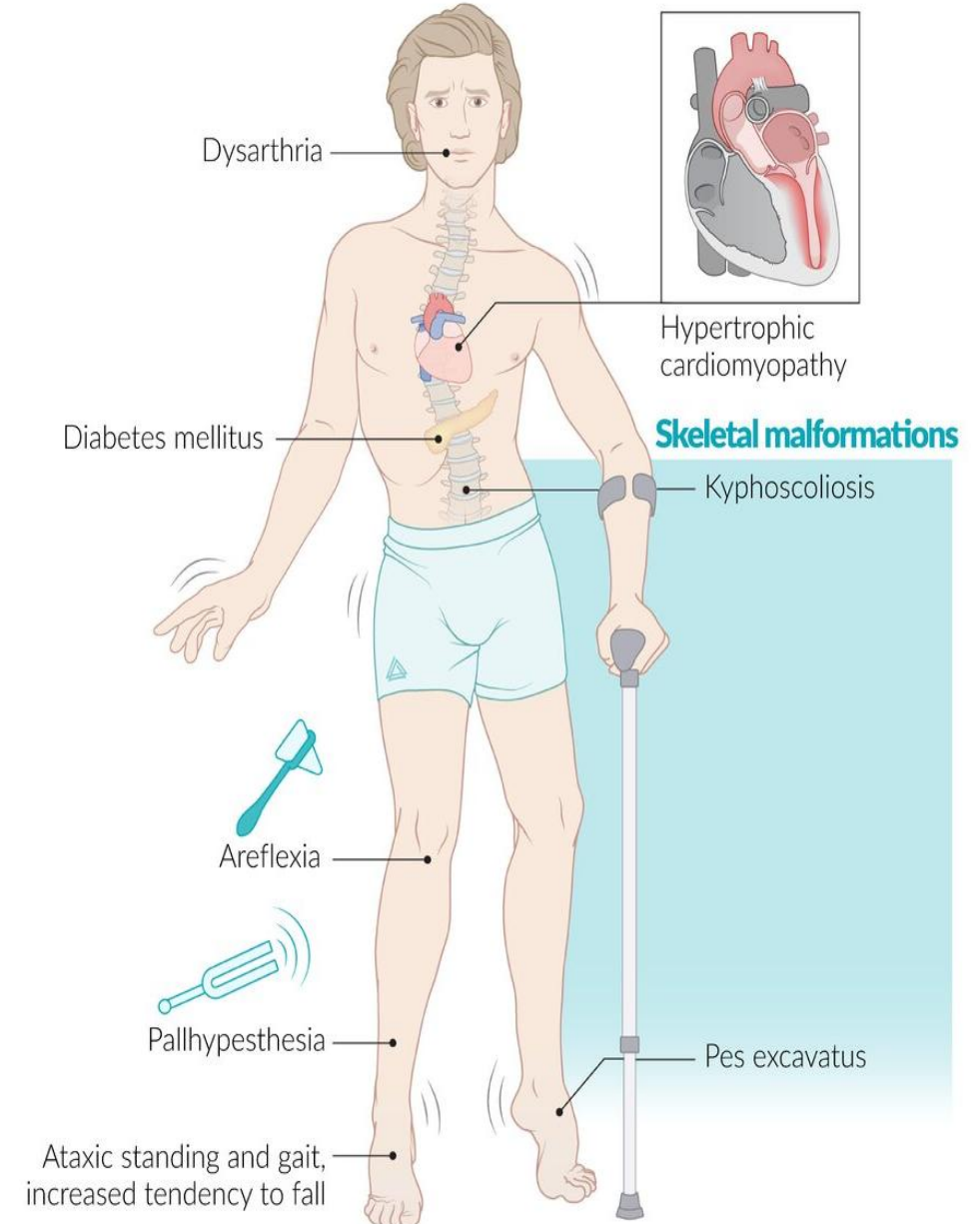


B - First physician visited

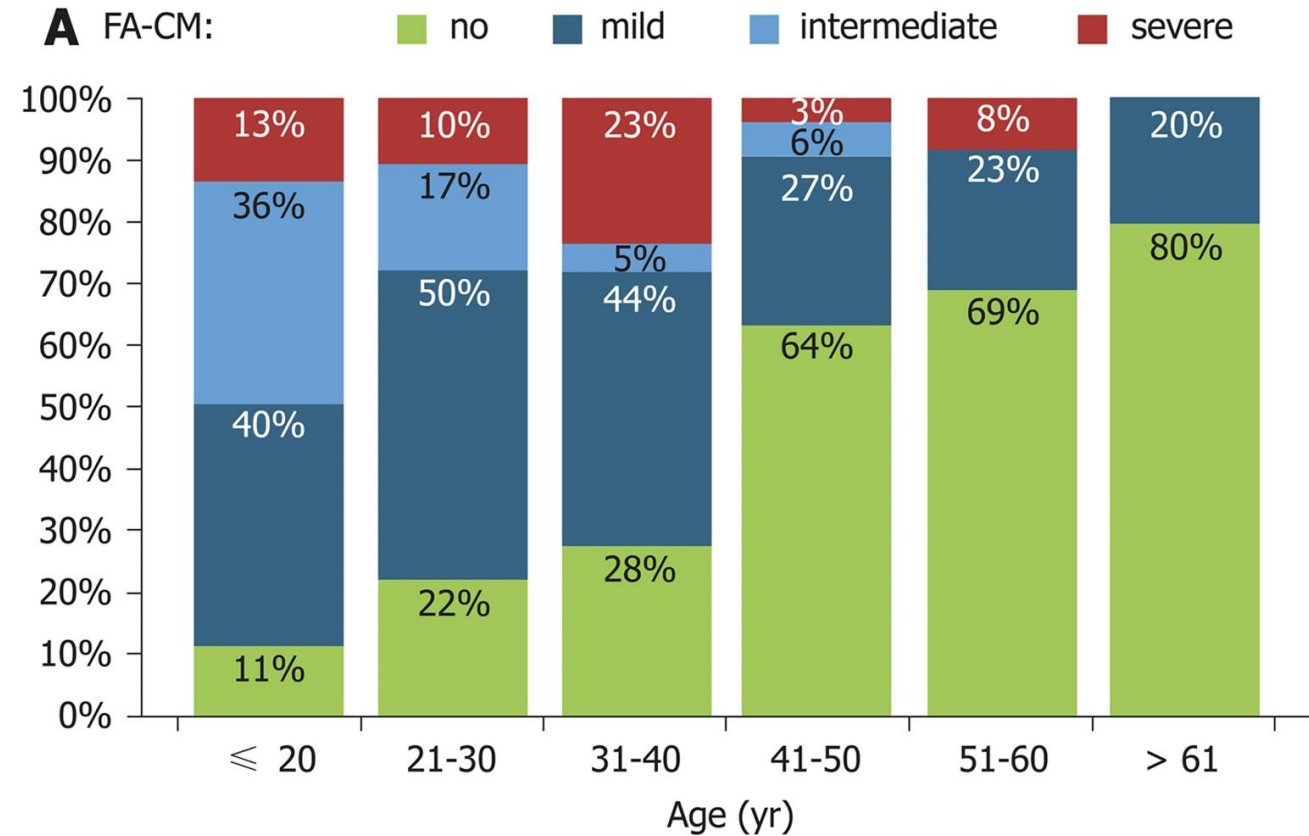
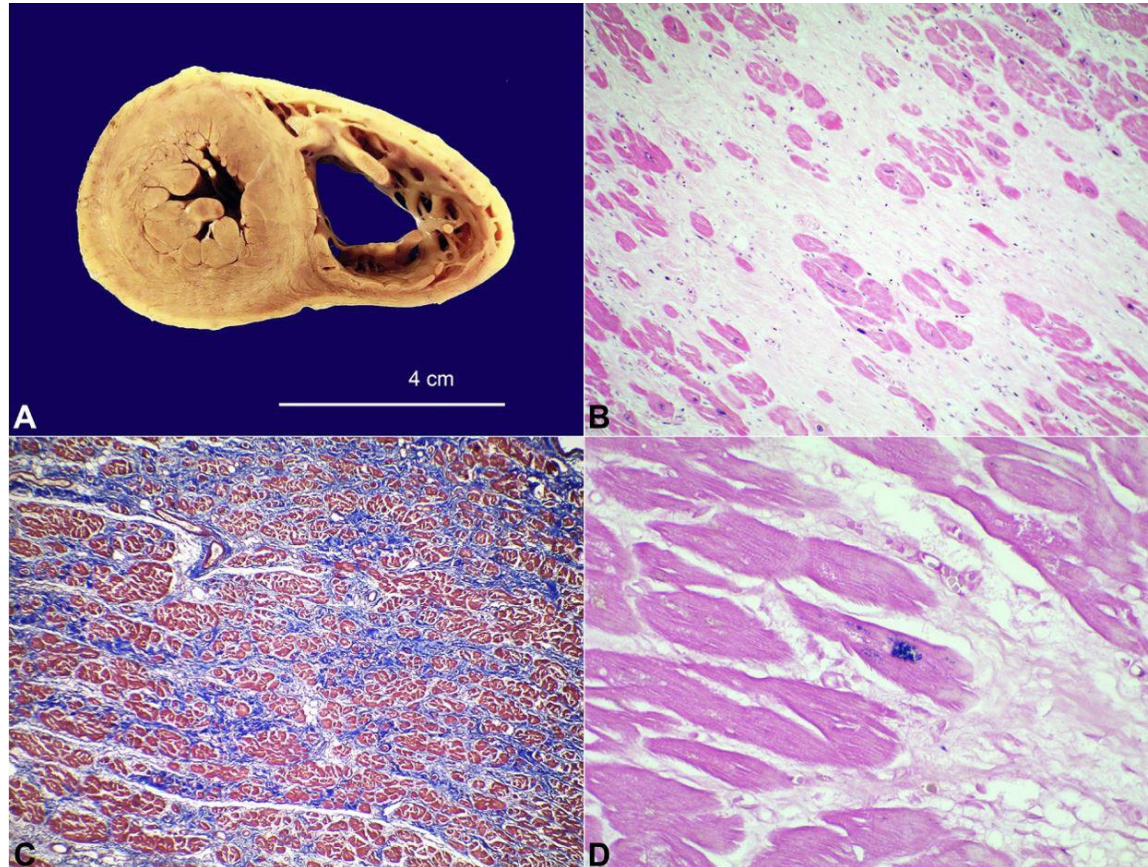


Ataxia de Friedreich: Características-chave

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Ataxia de Friedreich: Dano cardíaco

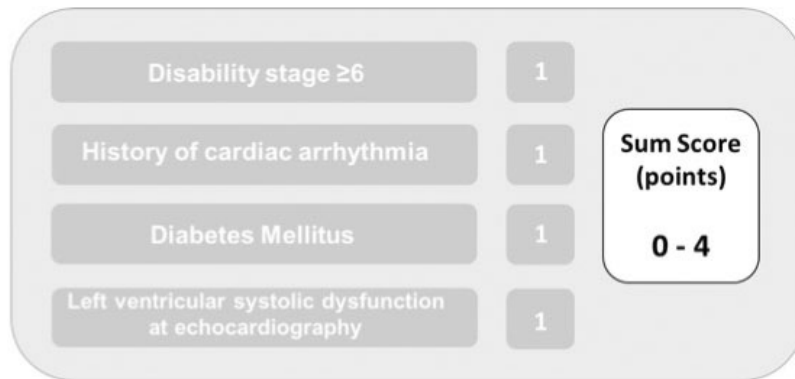


História natural: CM Hipertrofica → CM Dilatada

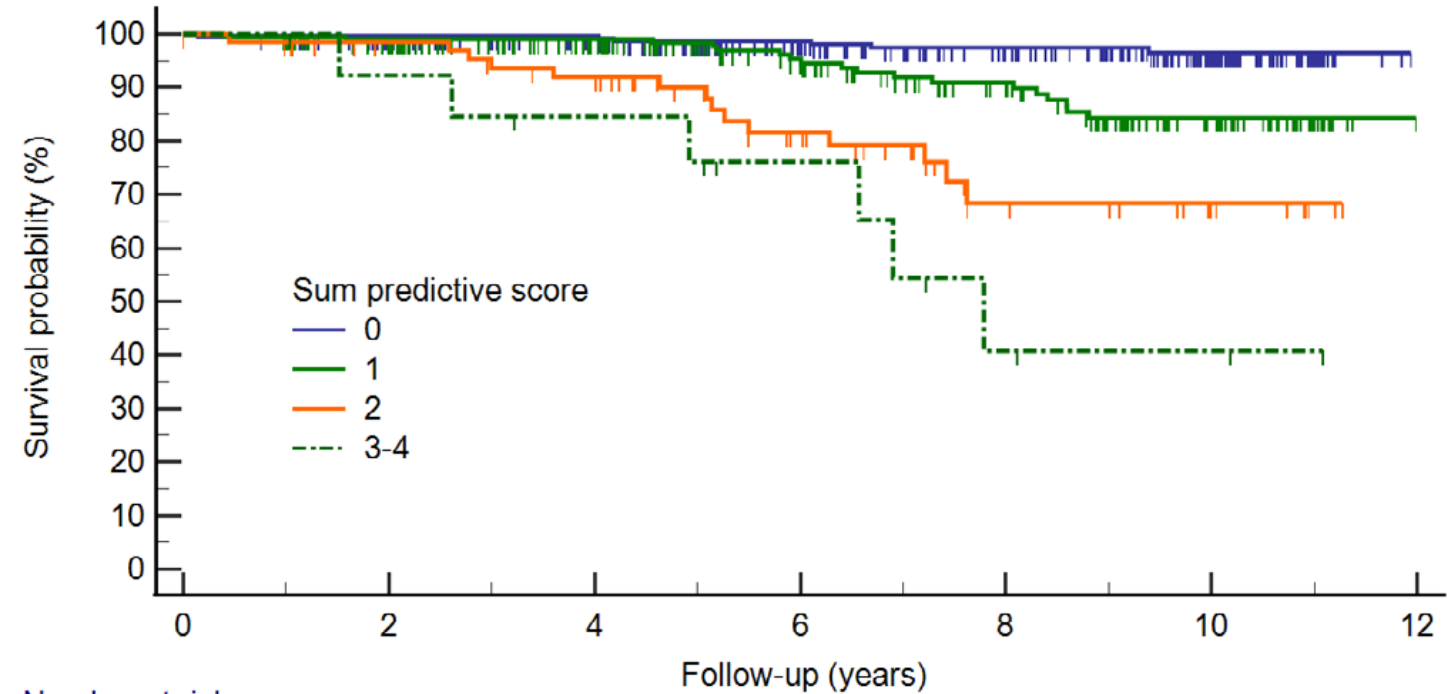
Ataxia de Friedreich: Sobrevida

Sobrevida média: 39 ± 14 anos

Causas : Cardíacas e
respiratórias



Sum score	10-years cumulative survival rate	95% CI
0	96.4%	93.4%-99.4%
1	84.3%	77.4%-91.3%
2	69.6%	55.4%-83.8%
≥ 3	42.4%	10.7%-74.1%



Number at risk

Group: 0

277 249 219 170 131 68 0

Group: 1

223 201 163 121 88 45 0

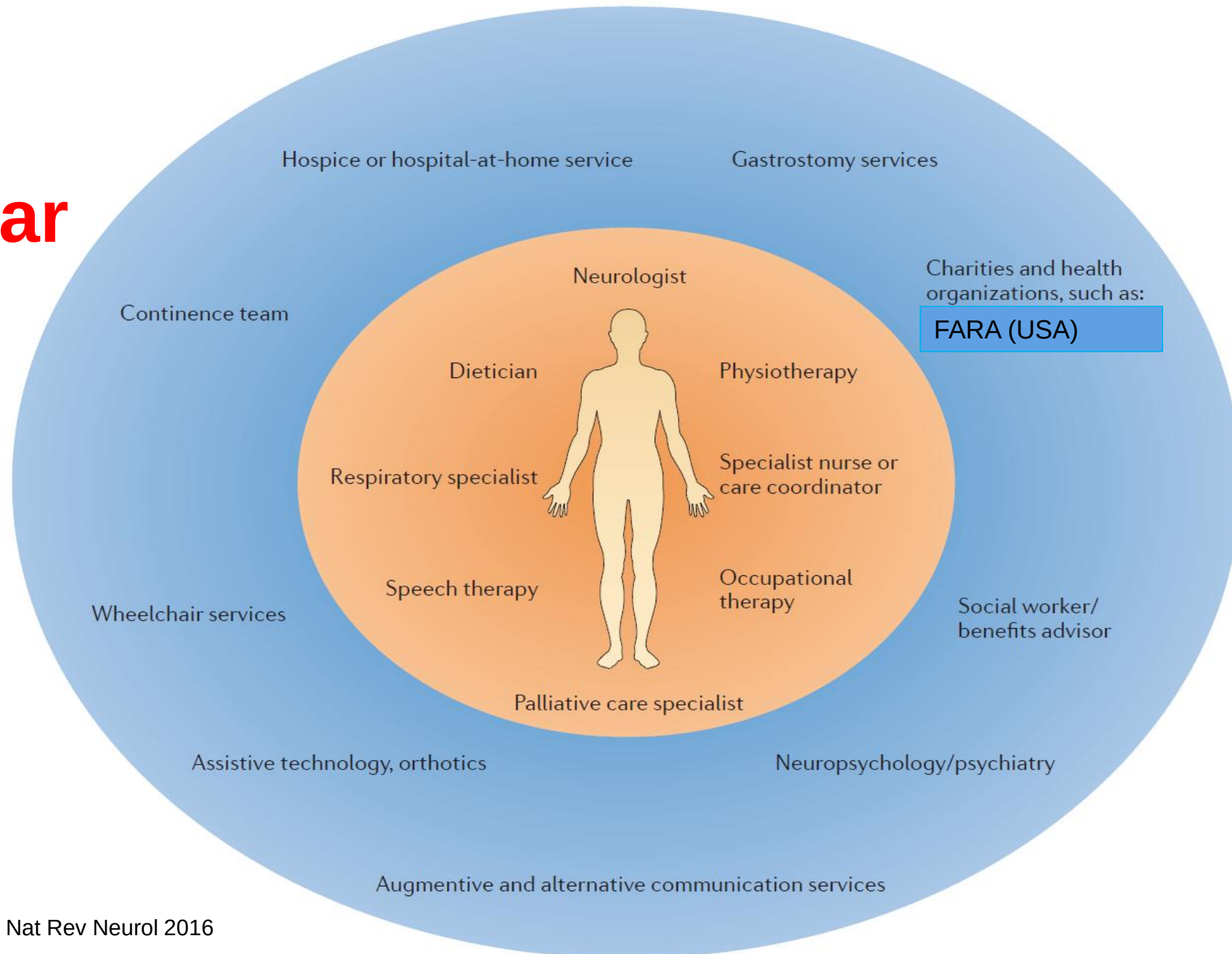
Group: 2

68 61 55 36 16 7 0

Group: 3-4

14 12 10 7 3 2 0

Cuidado multidisciplinar



FRIEDREICH'S ATAXIA TREATMENT PIPELINE

