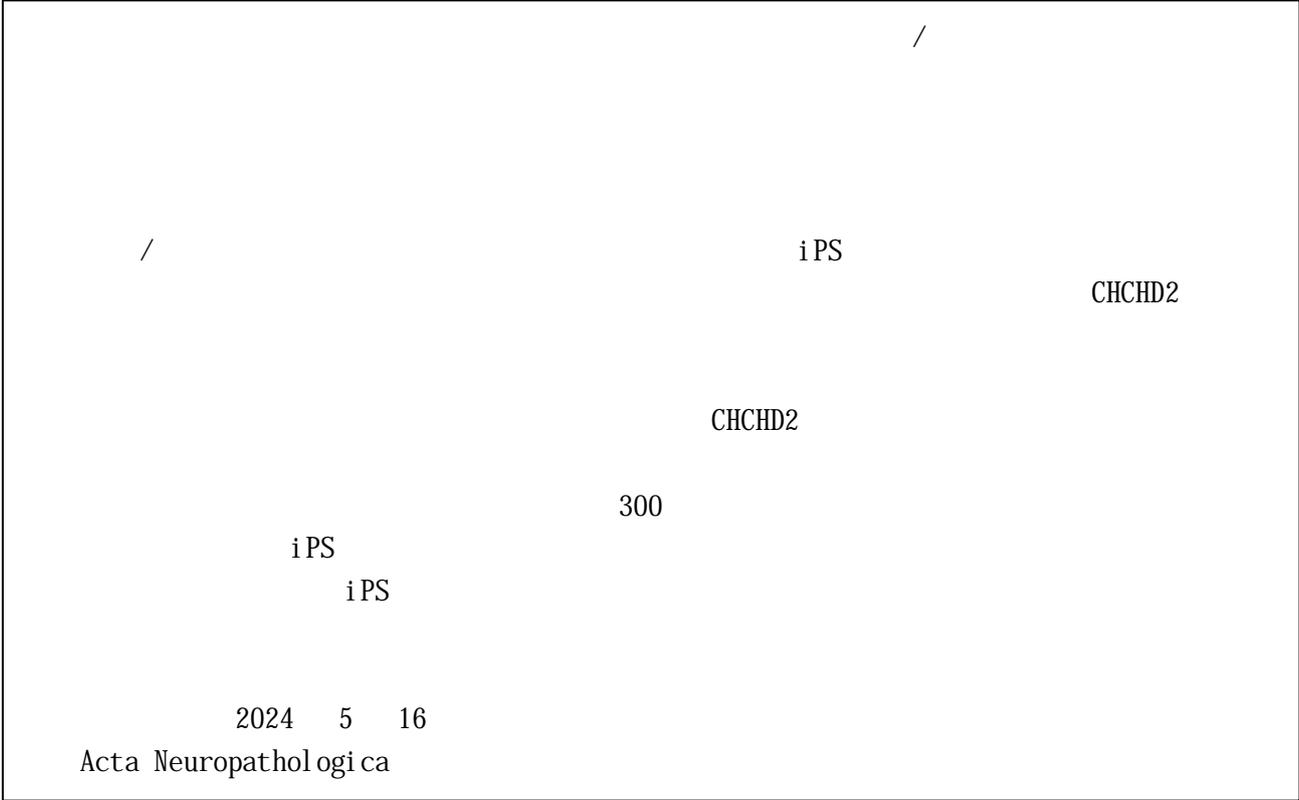


Kii ALS/PDC

iPS



1 _____

/ ALS Kii ALS/PDC

iPS

CHCHD2

CHCHD2

CHCHD2

2

ALS

ALS

ALS

ALS

ALS

Alzheimer

NFT

ALS

1980

ALS

1990

ALS

PDC

PDC

ALS

PDC

ALS/PDC

300

Kuzuhara S, et al. Ann Neurol 2001, Kuzuhara S.

Rinsho Shinkeigaku 2021

iPS

Fujimori K et al. Nat Med 2018, Okano H and Morimoto S. Cell Stem Cell 2022, Morimoto S, et al. Cell Stem Cell 2023

3

Kii

ALS/PDC

iPS

Kii ALS/PDC

iPS

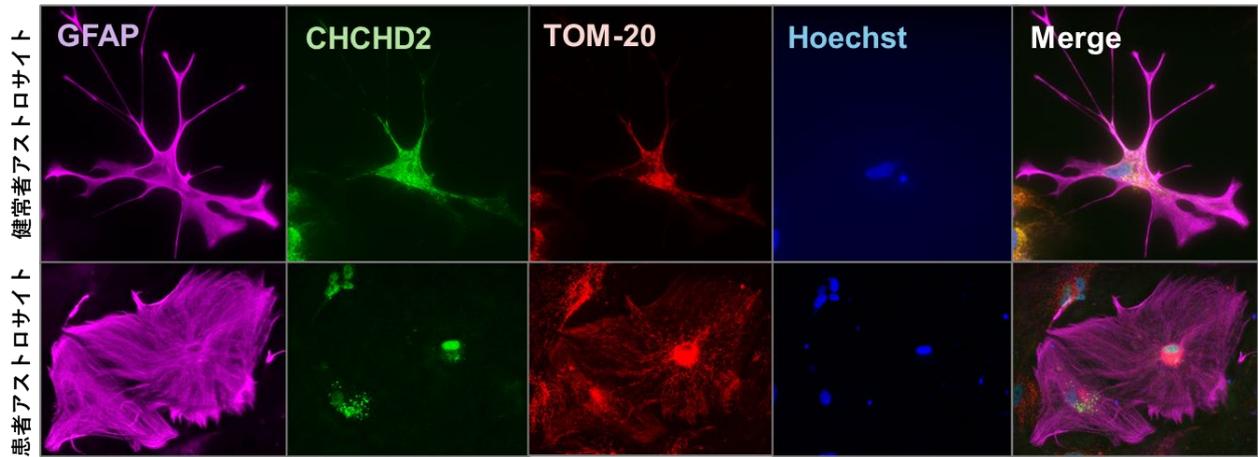
RNA-seq

CHCHD2

CHCHD2

Elamipretide

CHCHD2



iPS

CHCHD2

iPS

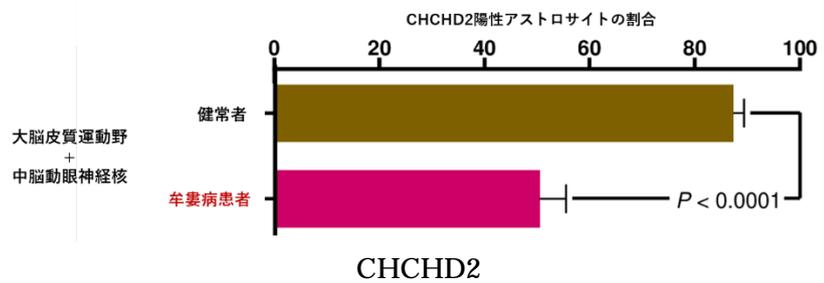
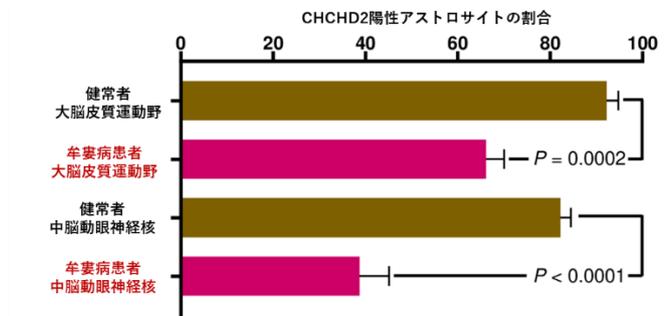
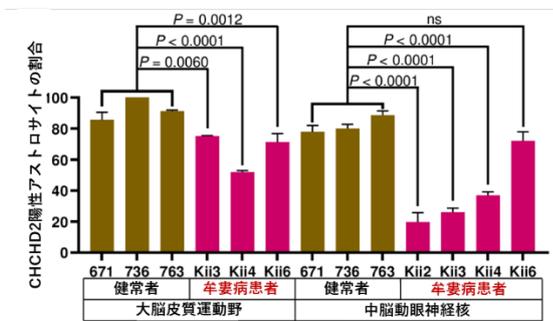
GFAP
GFAP
TOM-20

CHCHD2

iPS

CHCHD2

ALS Kii ALS



4

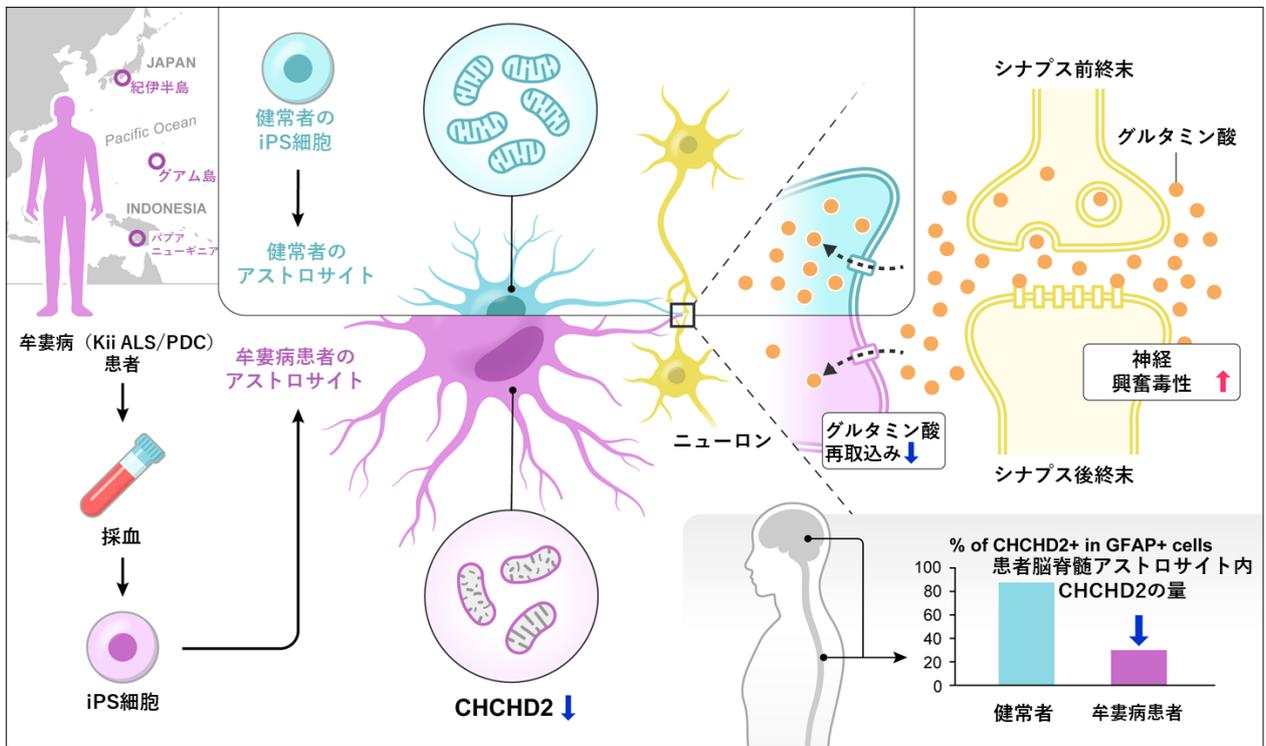
iPS

iPS

Kii ALS/PDC

iPS
iPS
Kii ALS/PDC

CHCHD2



5. _____

AMED

iPS

iPS

(ALS)

(ALS)

iPS

1/2a

RNA

RNA

JSPS

RNA

JP20H00485, JP21F21410,

JP21H05278, JP22KF0333, JP22K07500, JP22K15736

ALS

ALS

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Aberrant CHCHD2-associated mitochondriopathy in Kii ALS/PDC astrocytes
ALS/PDC CHCHD2

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1 iPS 子 入 立 人

(human induced pluripotent stem cell)

2

3 CHCHD2

ATP

4 iPS

iPS

5

2-4

6

7

ATP

8 Alzheimer

NFT

9

10 RNA-seq

mRNA

11

12

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