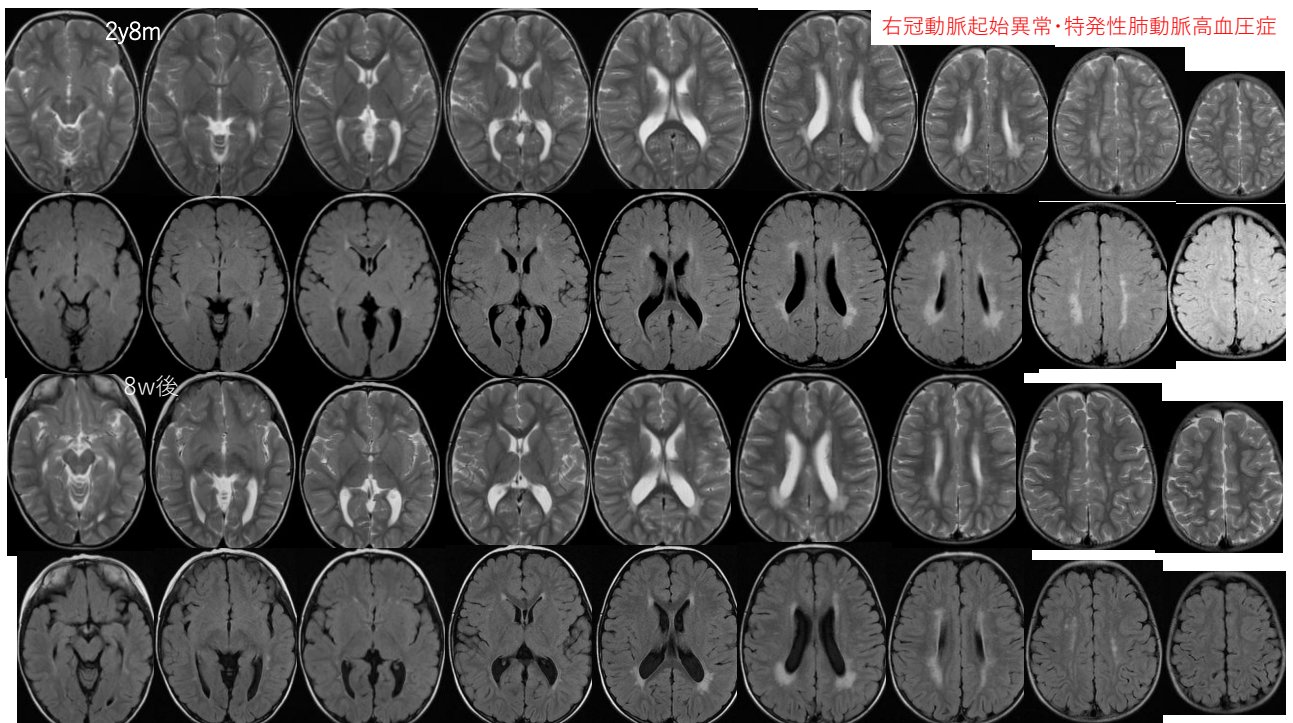


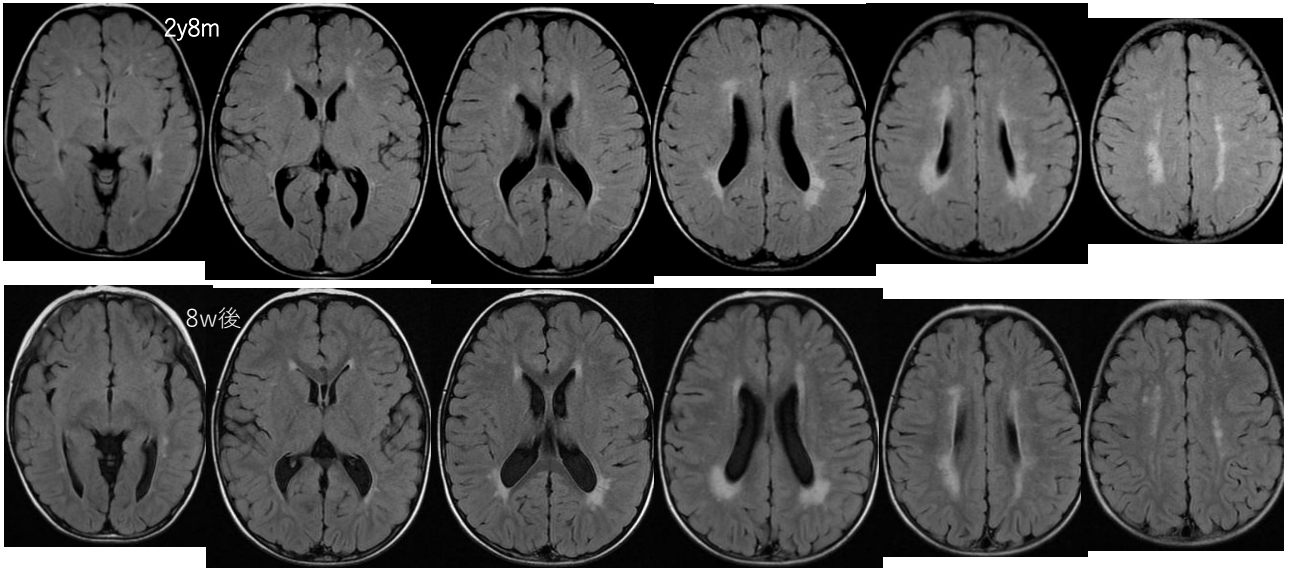
先天性脳障害の白質障害

横地健治

1

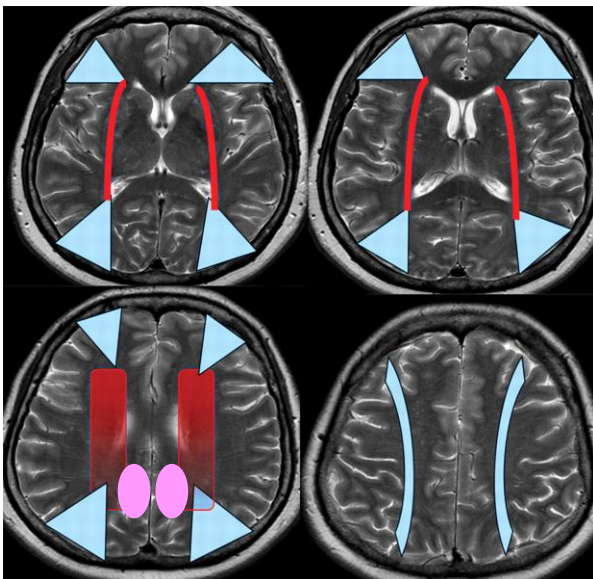


2



- ・ Iris signのある側脳室周囲病変
- ・ 半卵円中心の斑点病変 → Deep medullary vein thrombosis
- けいれん重積・心停止
- × 先天性白質障害 × 白質変性症 × 内部型境界境界域梗塞 ○ 静脈梗塞

3



Color overlays on axial T2-weighted magnetic resonance (MR) images of normal cerebrum show probable locations of external (blue) and internal (red) border zone infarcts.

The external or cortical border zones

- the **anterior** external border zones と paramedian white matter は、ACAとMCAの境界域
- the **posterior** external border zones (the parieto-occipital areas) は、MCAとPCAの境界域

● 大脳内側後方のACAとPCAの境界域は？

The internal or subcortical border zones

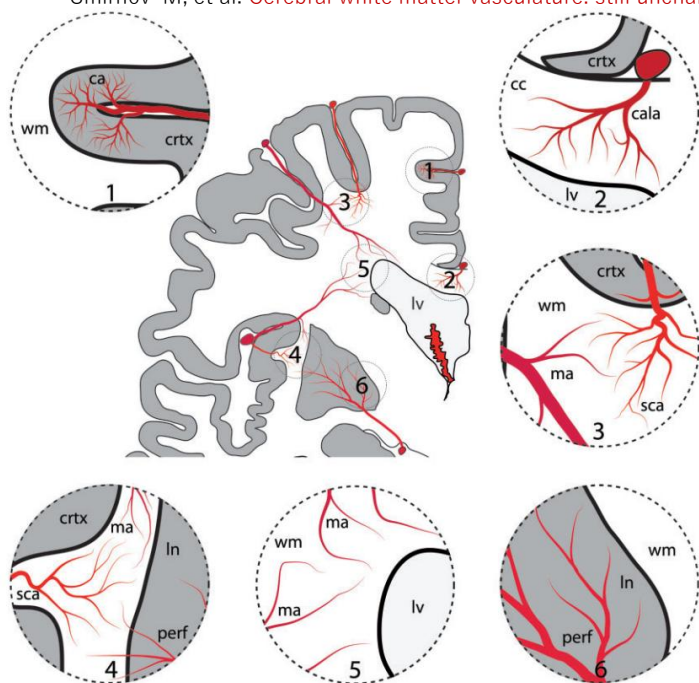
- the Heubner artery (長中心動脈(反回動脈)、ACA)、the lenticulostriate artery (MCA)、the anterior choroidal artery (内頸動脈) との境界域
- the lenticulostriate artery と MCA
- the lenticulostriate artery と ACA
- the Heubner artery と ACA
- the anterior choroidal artery と MCA
- the anterior choroidal artery と PCA

Rajiv Mangla, Balasubramanya Kolar, Jeevak Almast, Sven E. Ekholm. Border Zone Infarcts: Pathophysiologic and Imaging Characteristics.

RadioGraphics 2011

4

Smirnov M, et al. Cerebral white matter vasculature: still uncharted? Brain 2021;144:3561–75.



Type 1: Arterial vascularization of the cortex depends on short arterioles originating from the same pial vessels (single source juxtaposed (並列) supply).

Type 2: The corpus callosum is supplied by short arterioles originating from the pial network (single source juxtaposed supply).

Type 3: Short association U-fibers are supplied by subcortical arteries and earliest branches of medullary arteries (dual source interdigitated supply).

Type 4: The external capsule, claustrum and extreme capsule take up subcortical arteries, earliest branches of medullary arteries, and terminals of perforating arteries of the base of the brain (triple source interdigitated supply).

Type 5: The centrum semiovale is supplied by terminals of deep white matter arteries (single source juxtaposed supply).

Type 6: Vessels supplying the basal ganglia and the thalamus are long, perforating arterioles and arteries originating from the base of the brain (single source juxtaposed supply).

cc = corpus callosum; ctx = cortex; lv = lateral ventricle; ma = medullary artery; perf = perforating artery; sca = subcortical artery; wm = white matter.

Type 5~6 : 内部型脳低血流障害

5

Khalatbari H, et al. Deep medullary vein engorgement and superficial medullary vein engorgement: two patterns of perinatal venous stroke. Pediatr Radiol 2021;51:675–685.

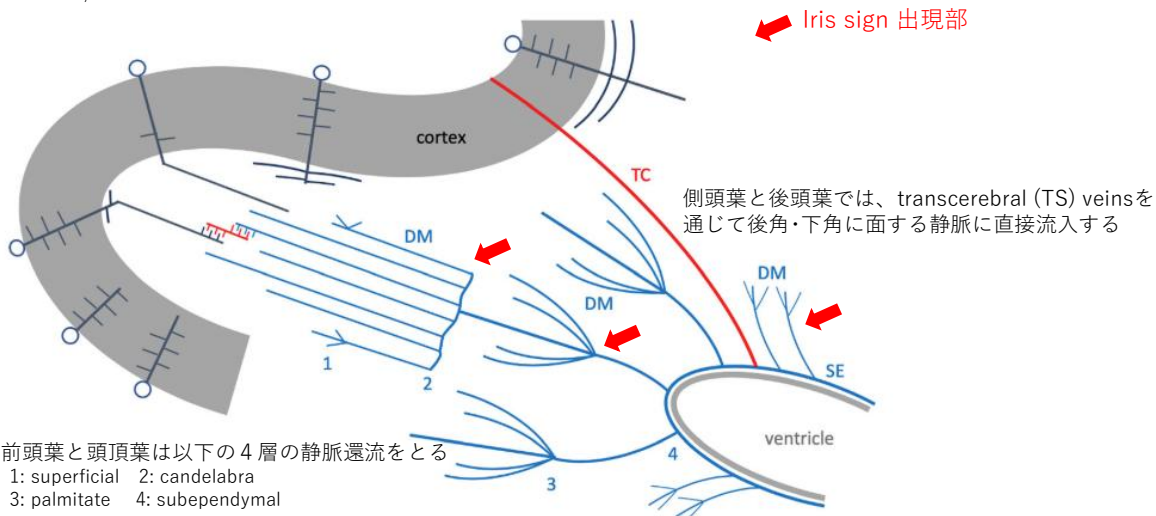


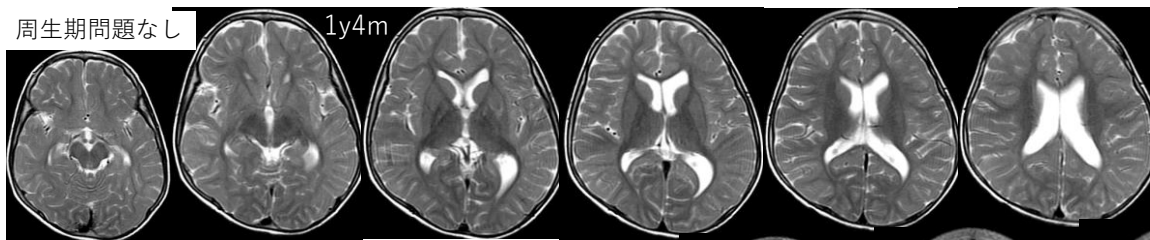
Fig. 2 The four zones of convergence of the deep medullary veins, the subependymal veins and the transcerebral veins. The deep medullary veins (DM) course centripetally to drain into the subependymal veins. Okudera et al. [13] demonstrated four zones of convergence in the frontal and parietal lobes by injecting barium into the internal jugular veins and obtaining radiographs of brain slices in cadavers. The zones of convergence from superficial to deep are the superficial (1), candelabra (2), palmitate (3) and subependymal (4) zones. In Zones 2 and 3 the medullary veins converge in characteristic configurations that resemble candelabras and palms, respectively. Transcerebral (TC) veins extend from the cortical surface to the subependymal veins. SE subependymal vein.

6

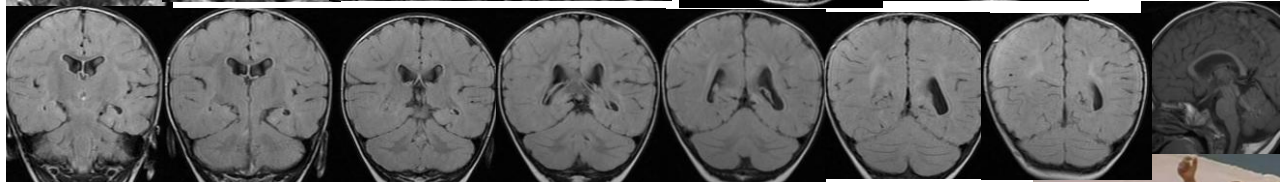
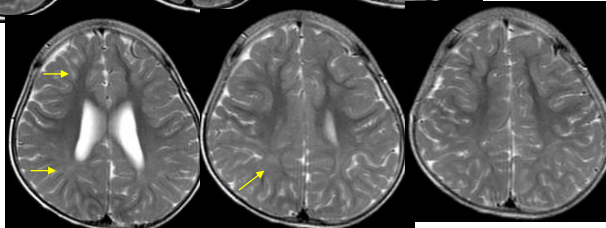
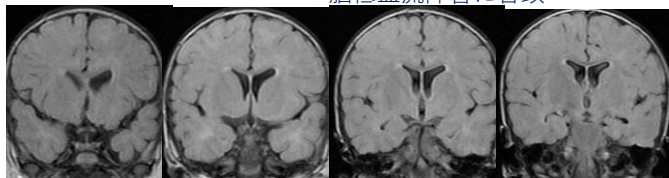
周生期間題なし

1y4m

座位1y9m
最重度ID



脳低血流障害に合致

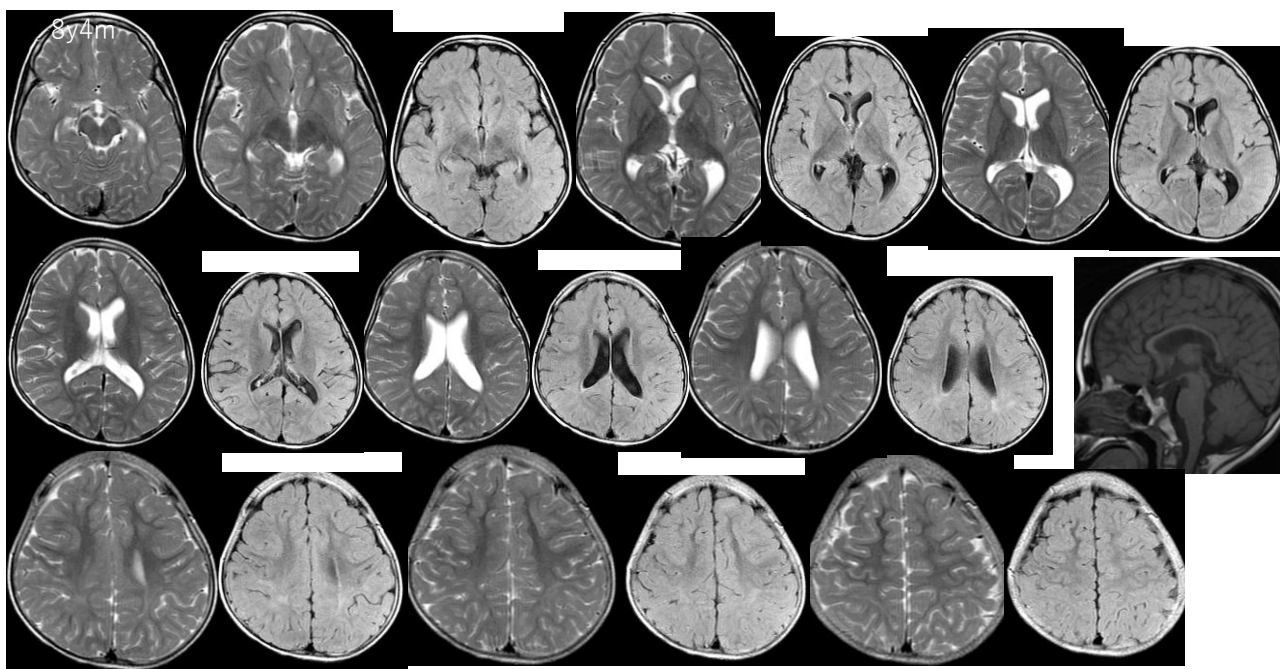


HIEの症候のない周生期脳低血流障害 or 先天性白質障害の後方優位型

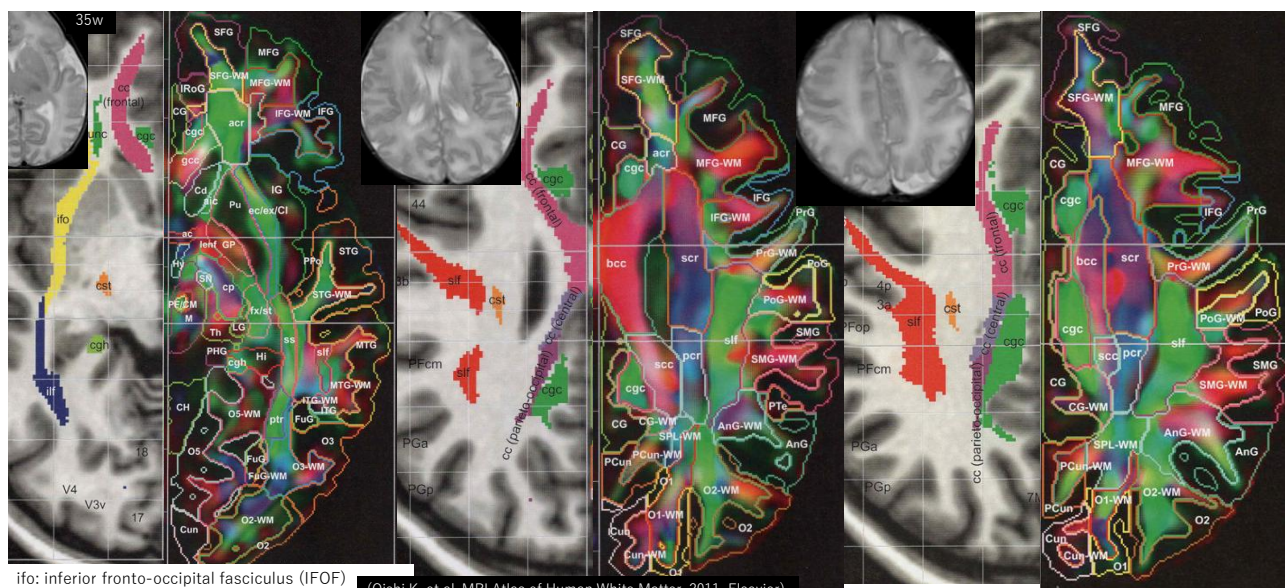
1y7m



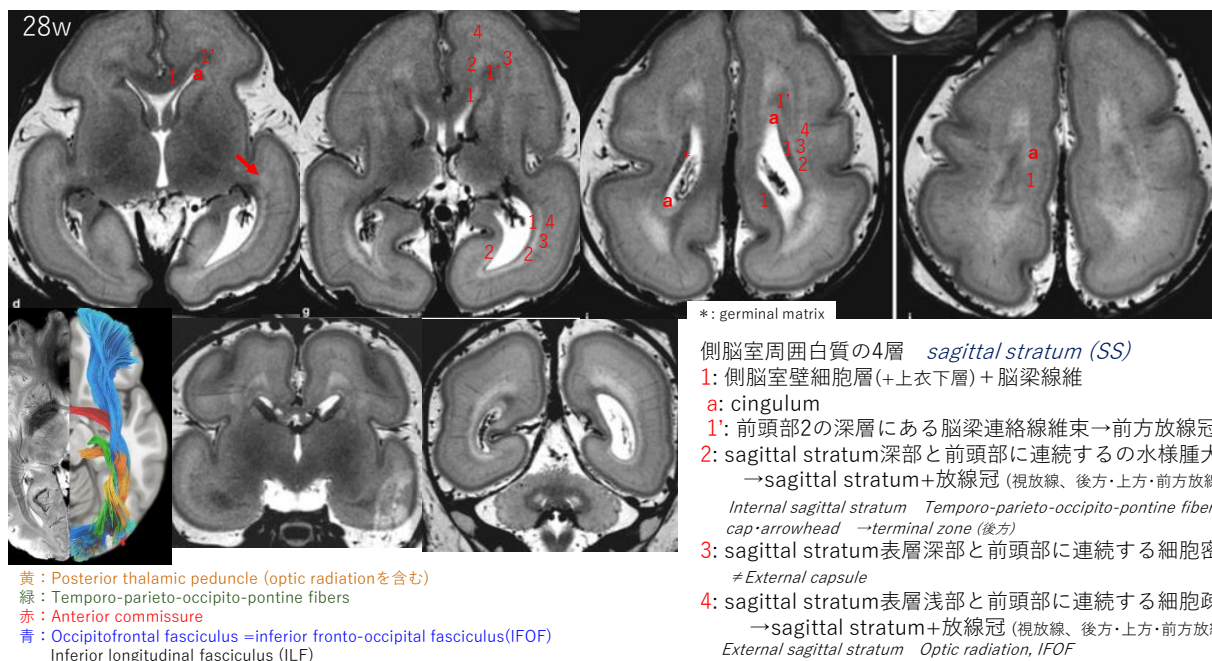
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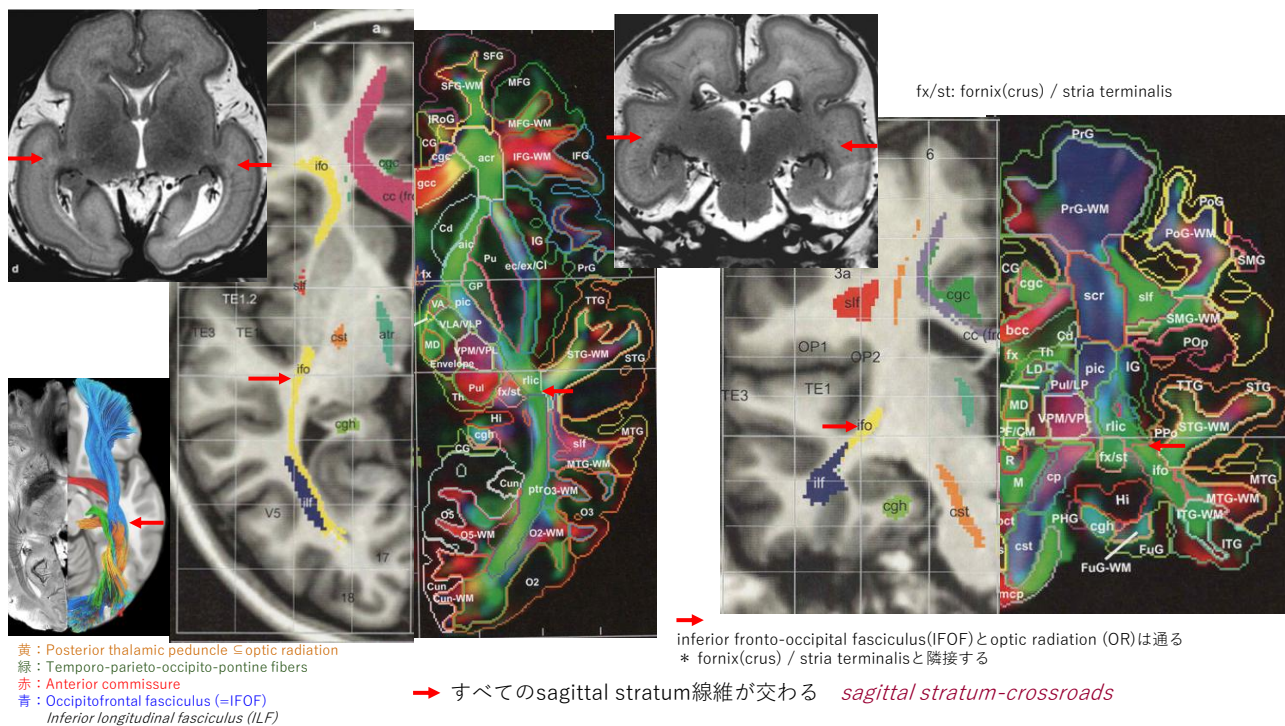
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9



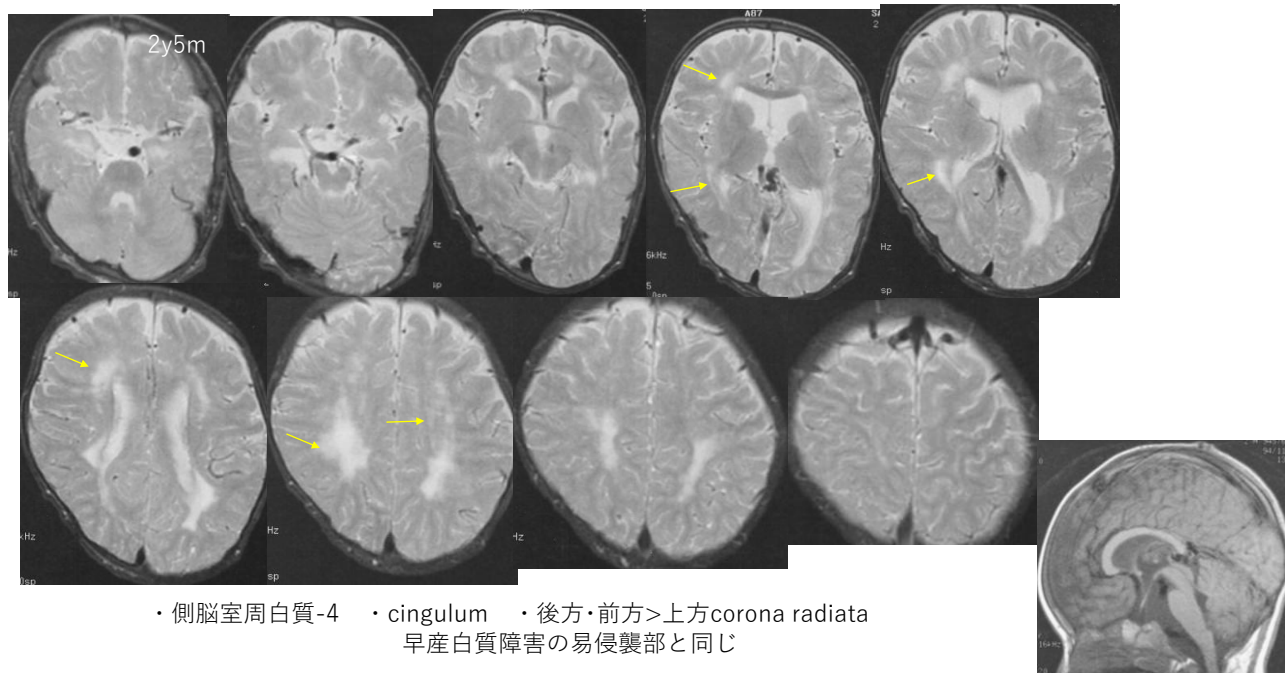
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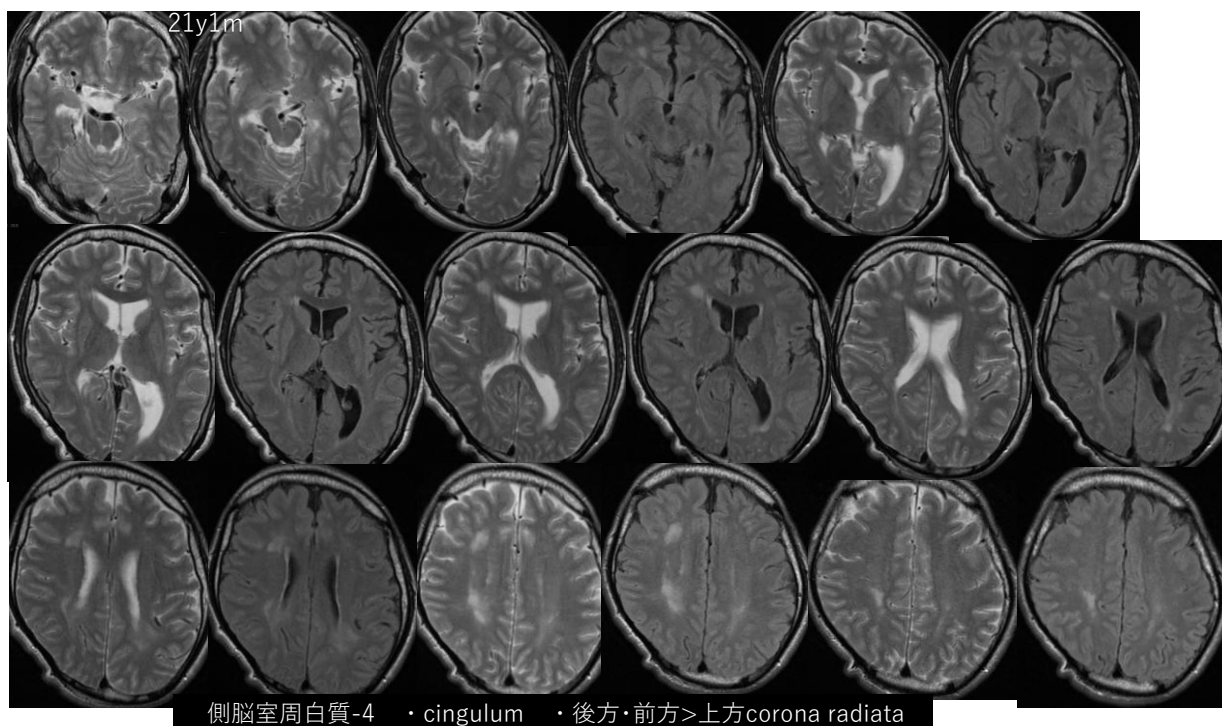
11

両側内頸動脈欠損

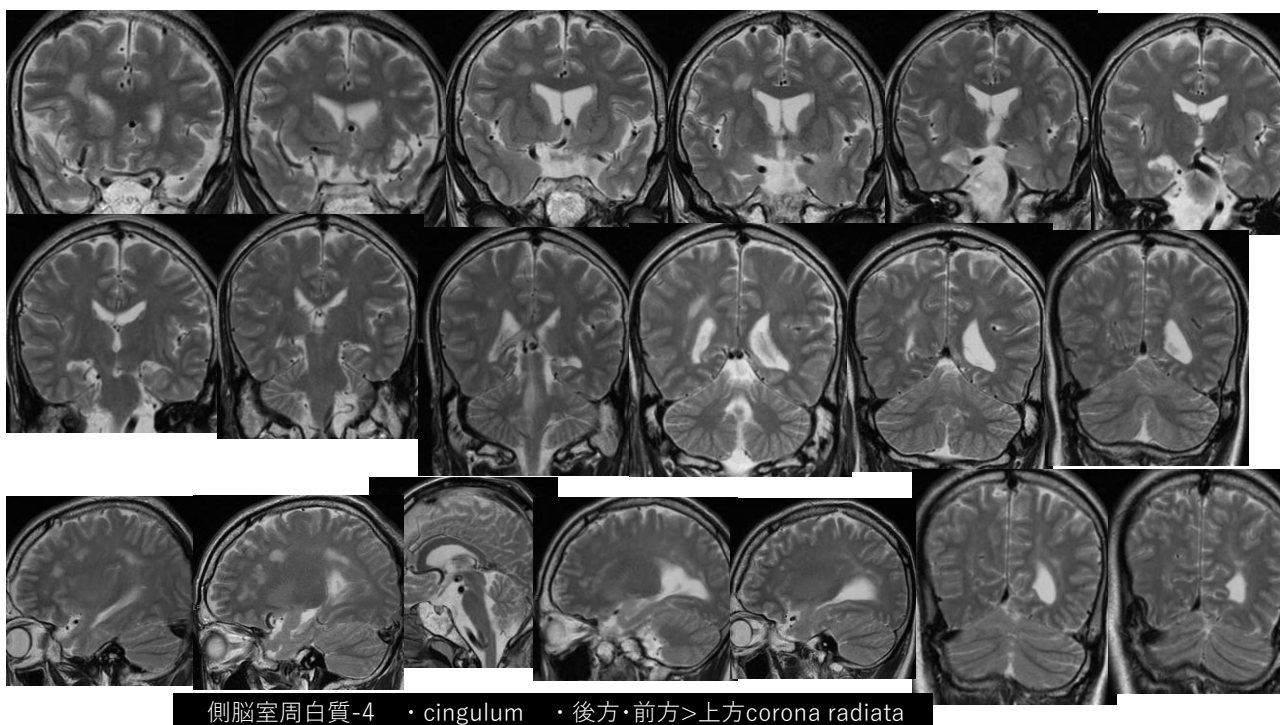
座位・いざり10y6m. 背臥位ブリッジ位が好む。有意な言語理解なし



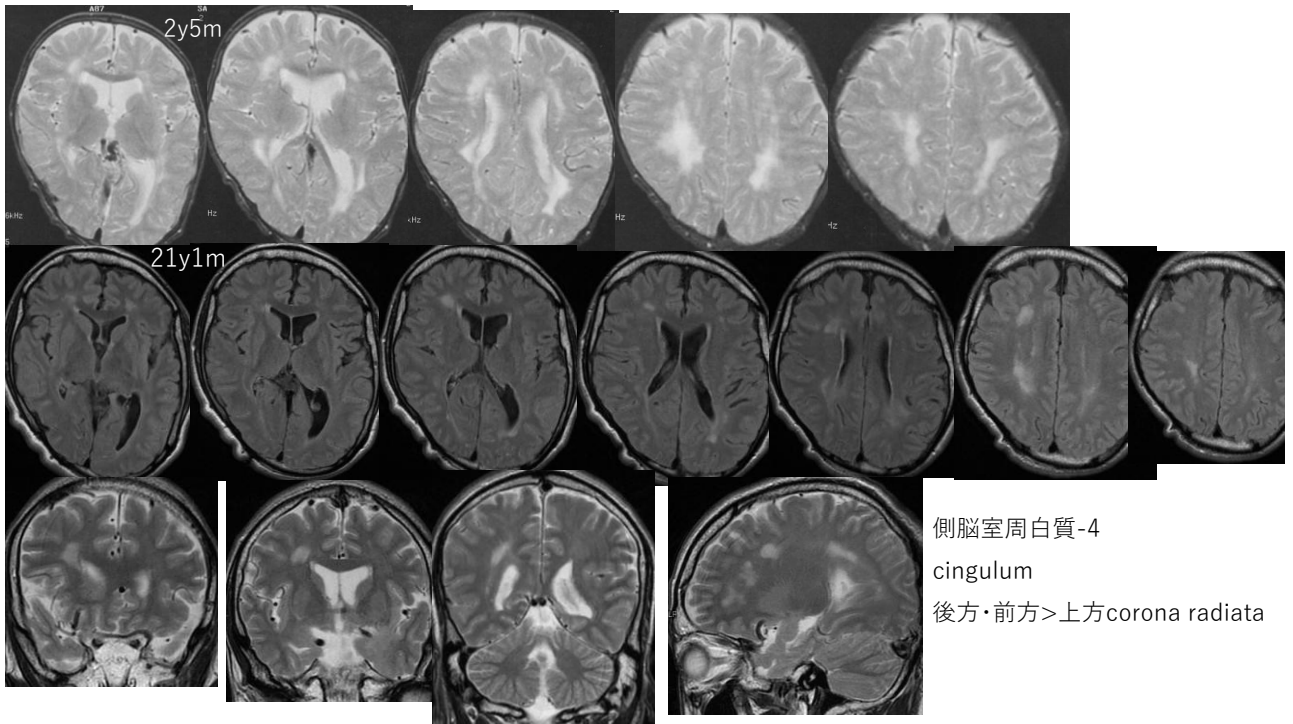
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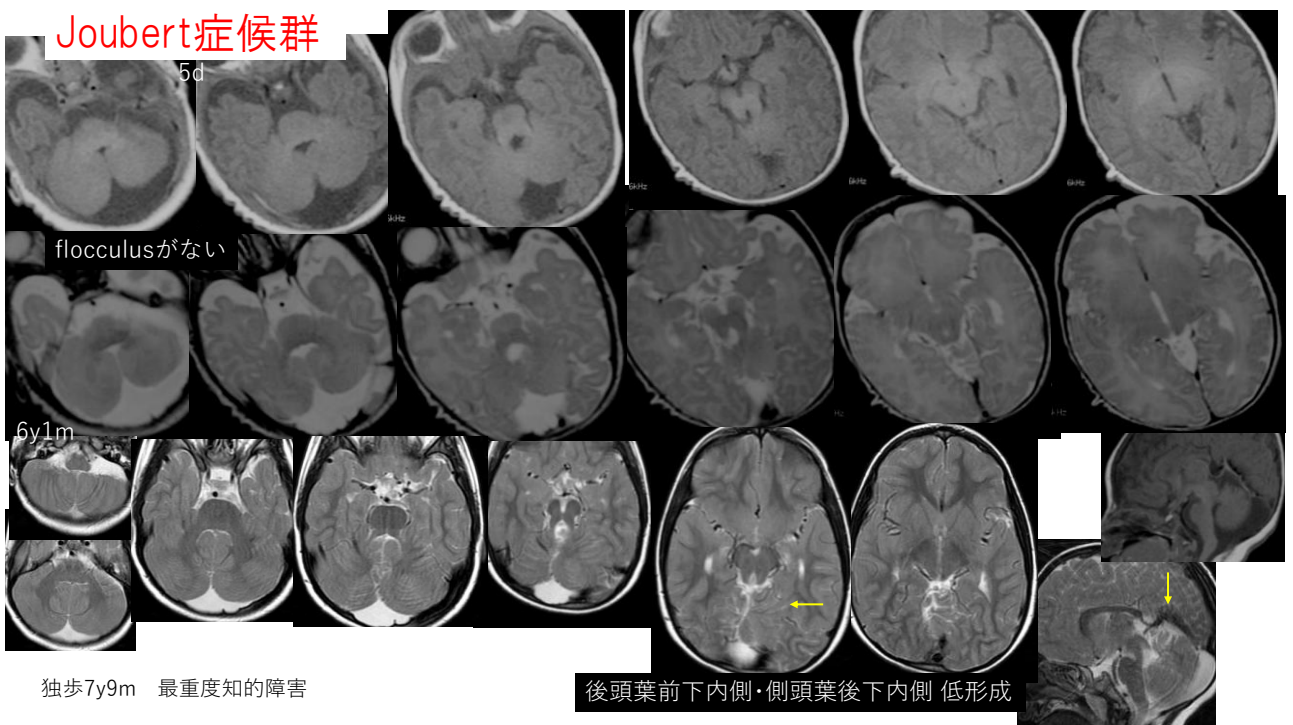
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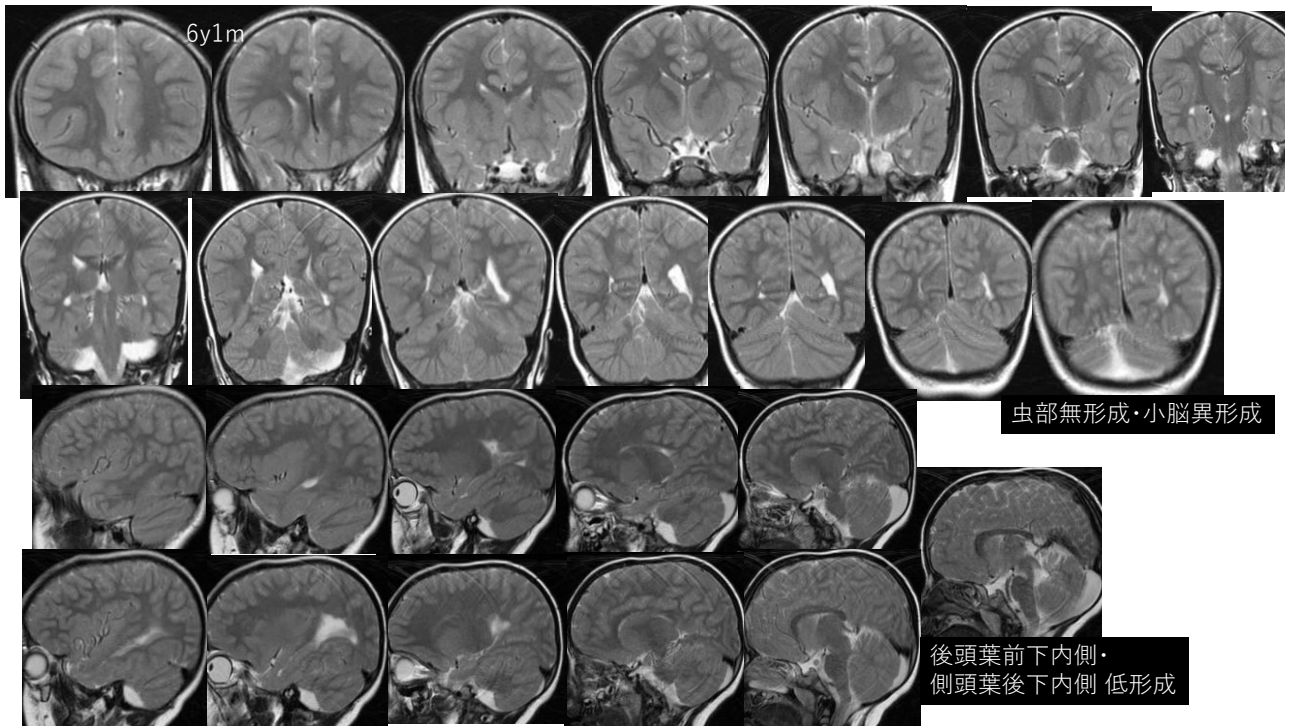
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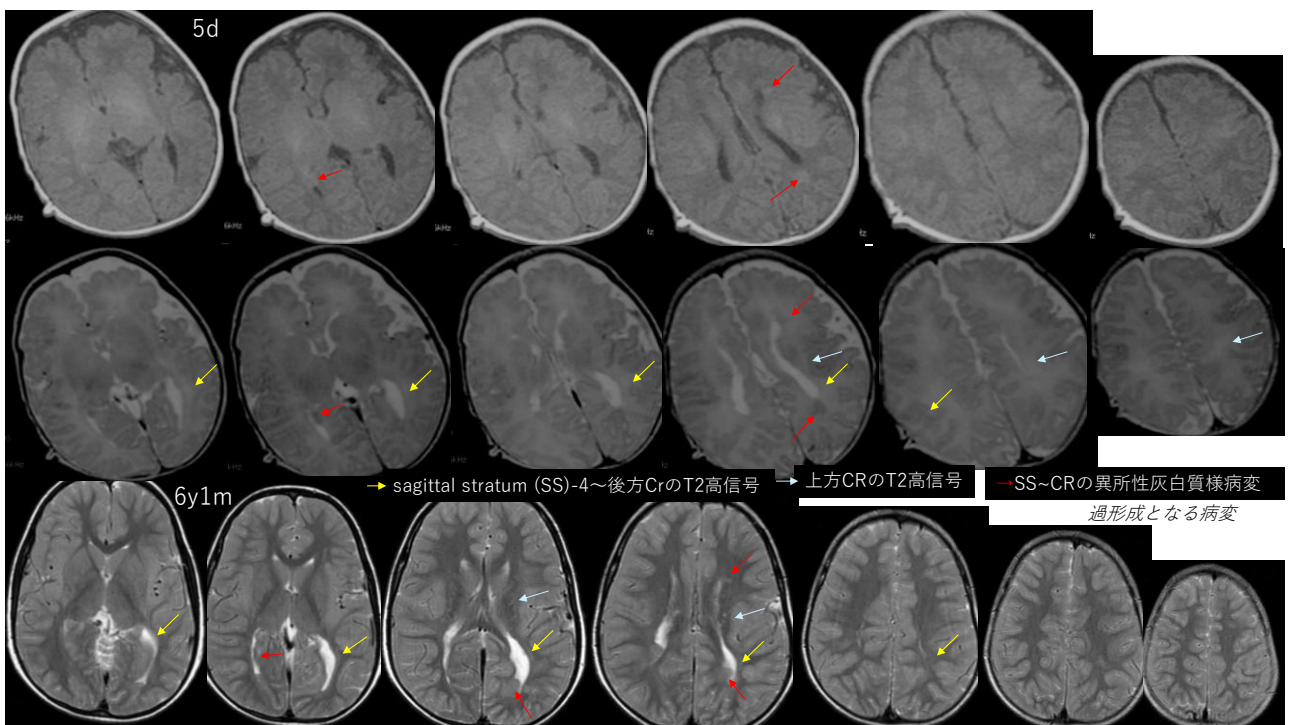
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16

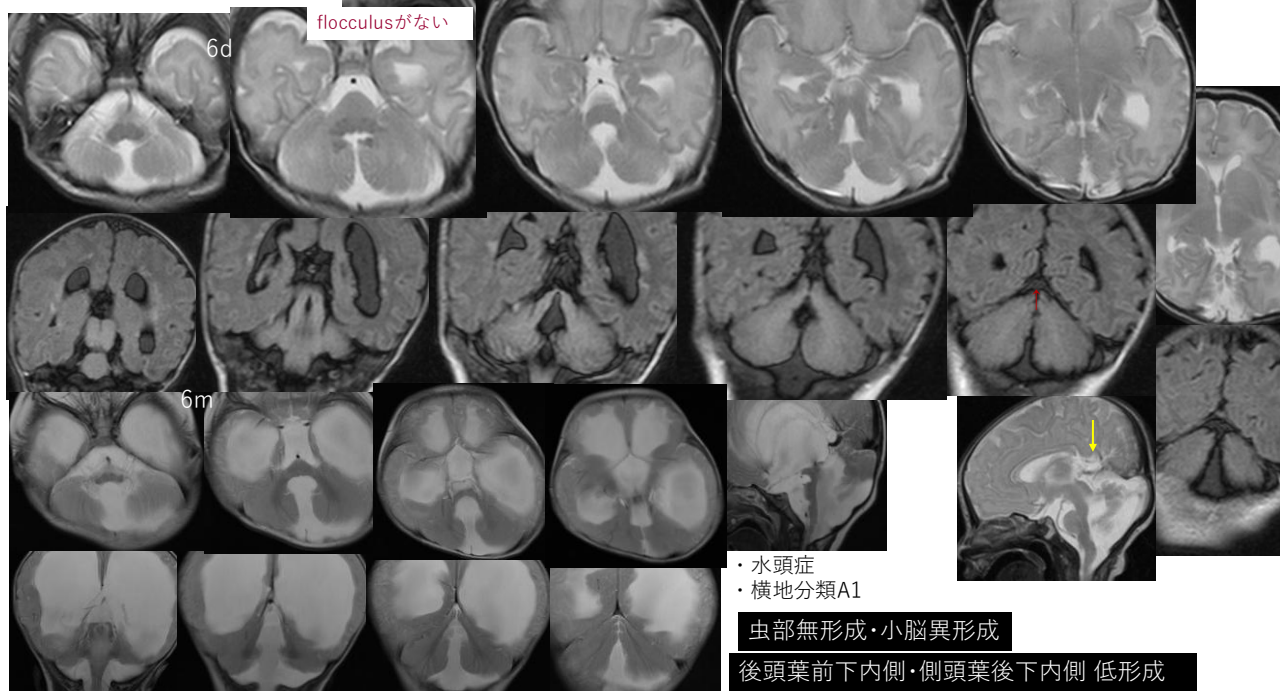


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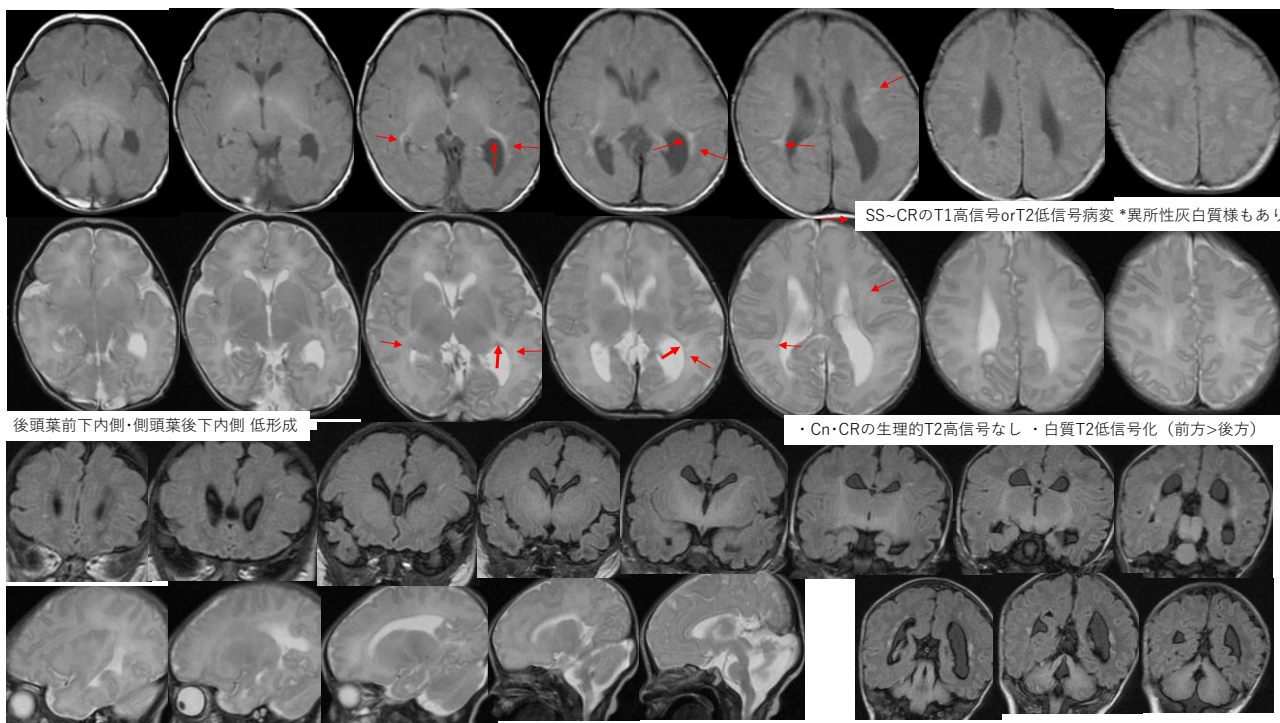


18

Joubert症候群・水頭症

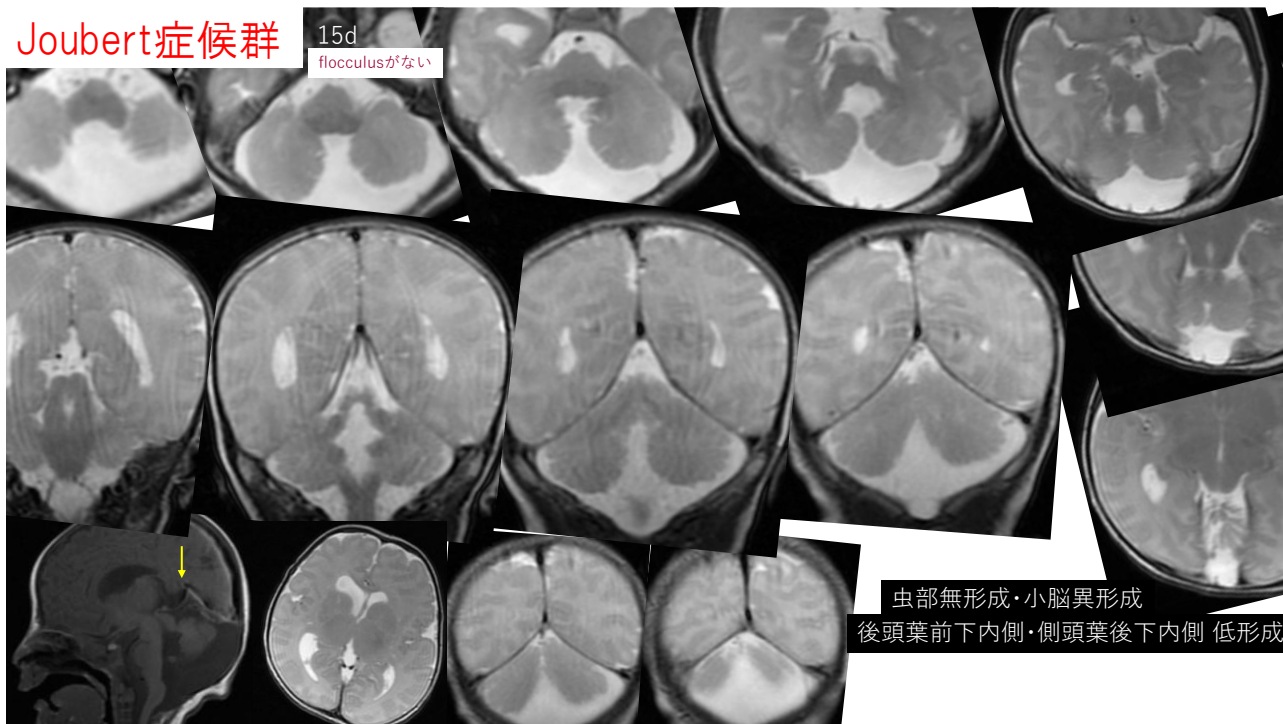


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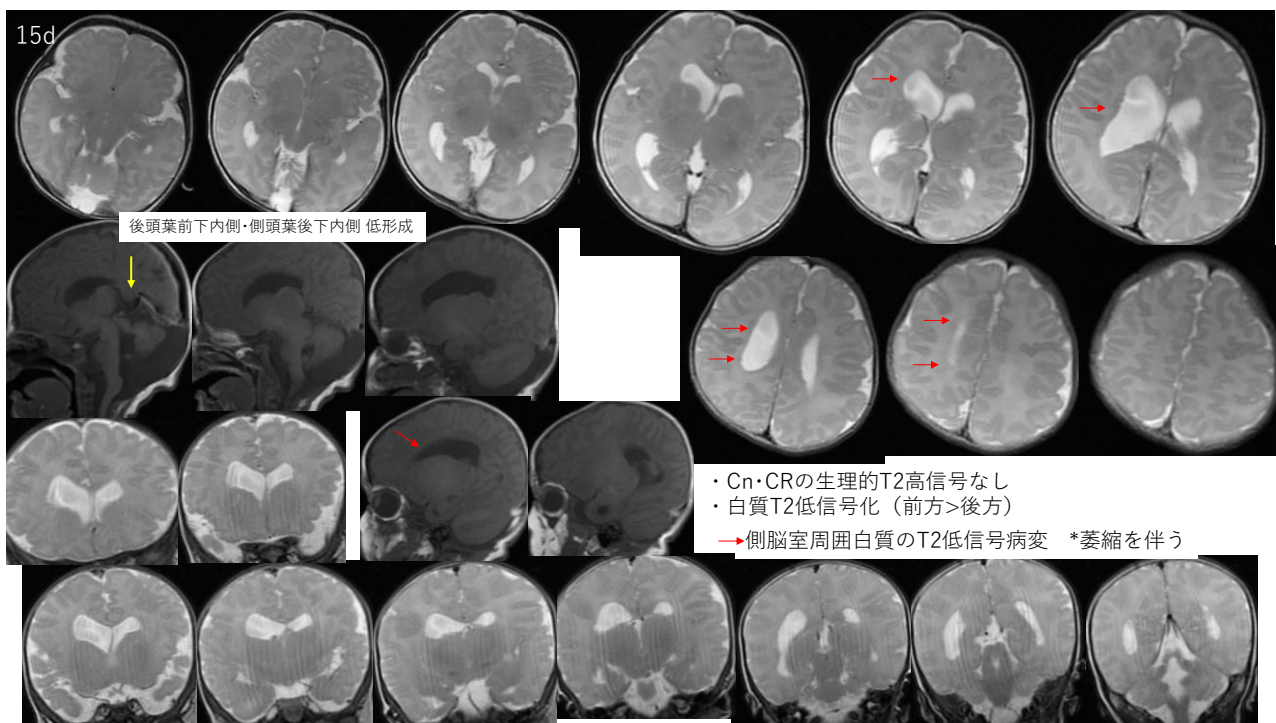


20

Joubert症候群

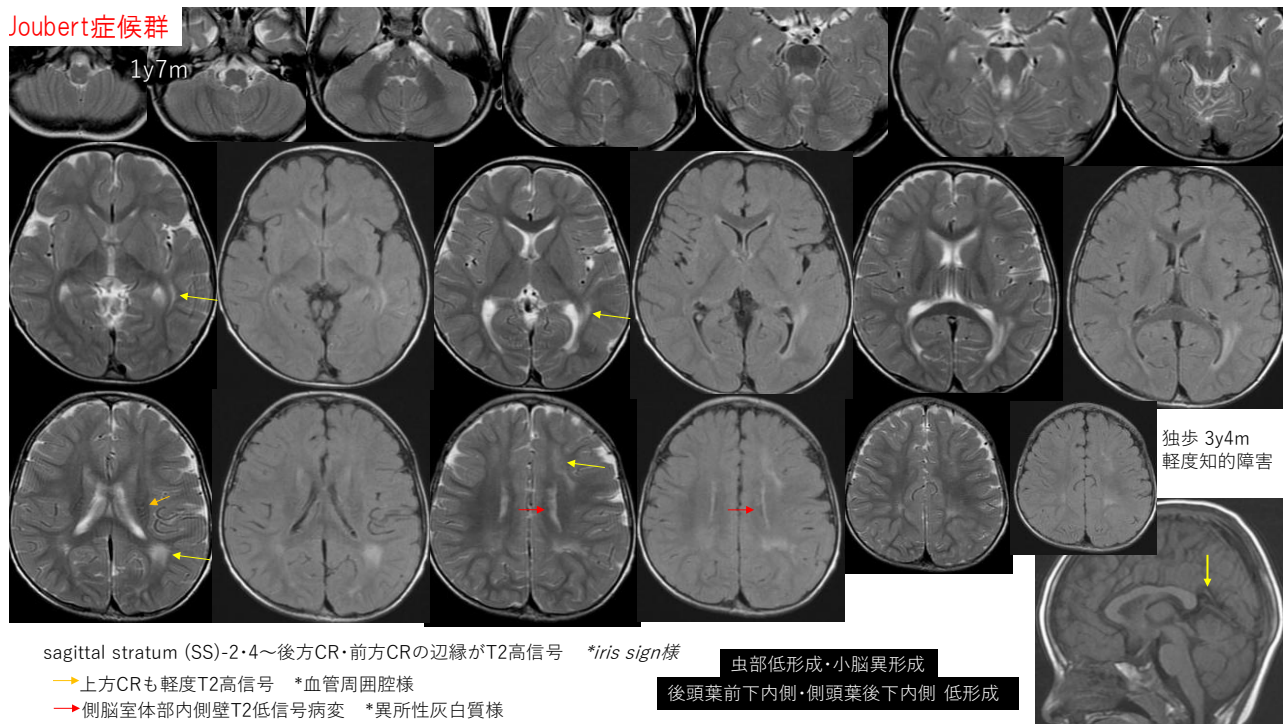


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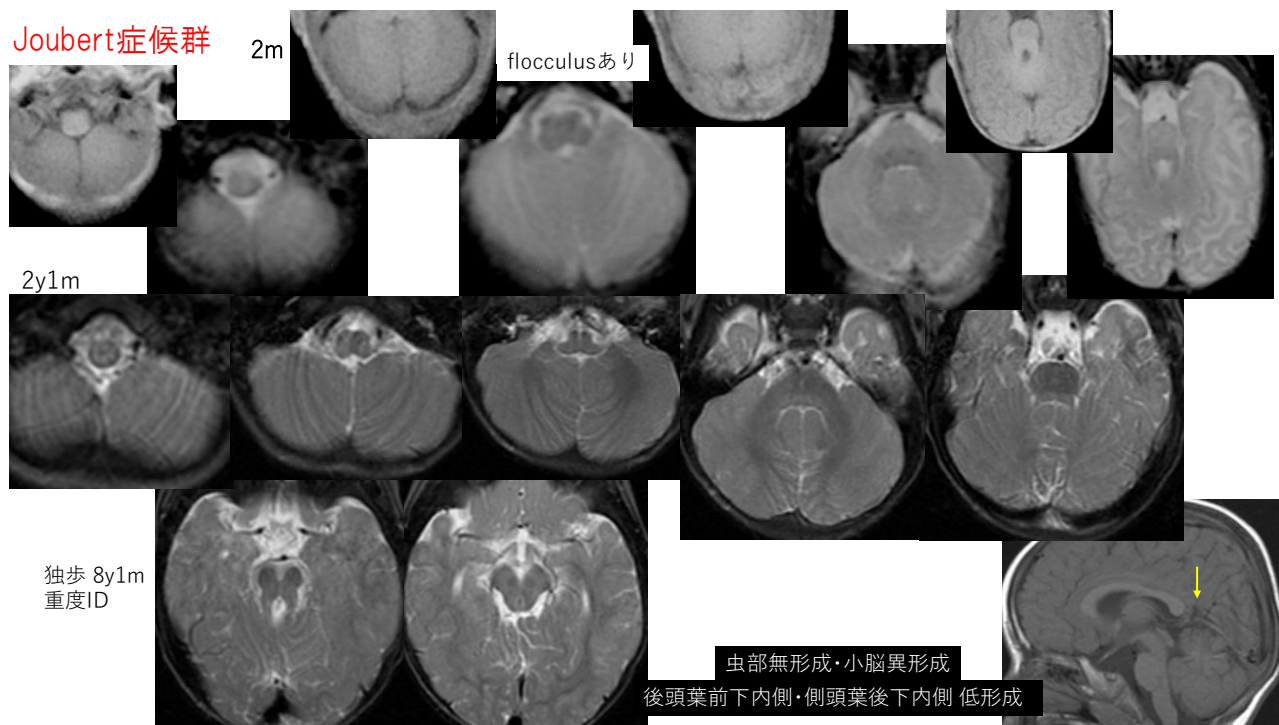
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Joubert症候群

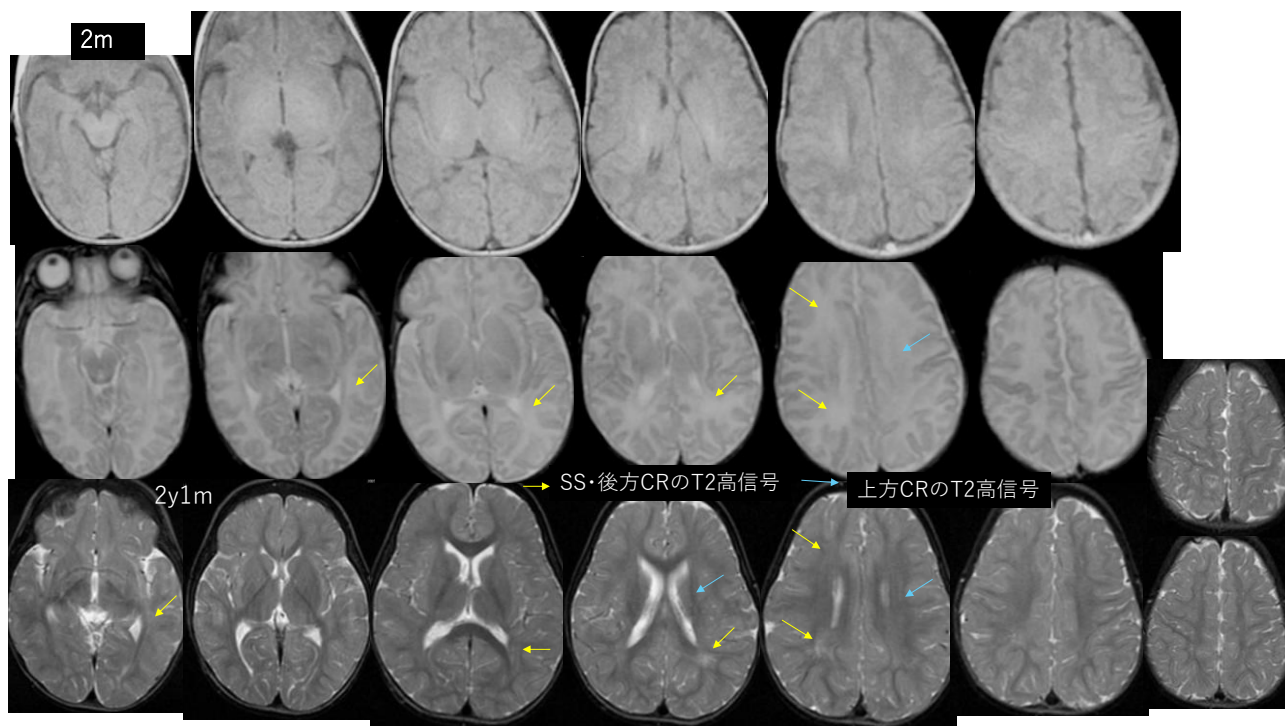


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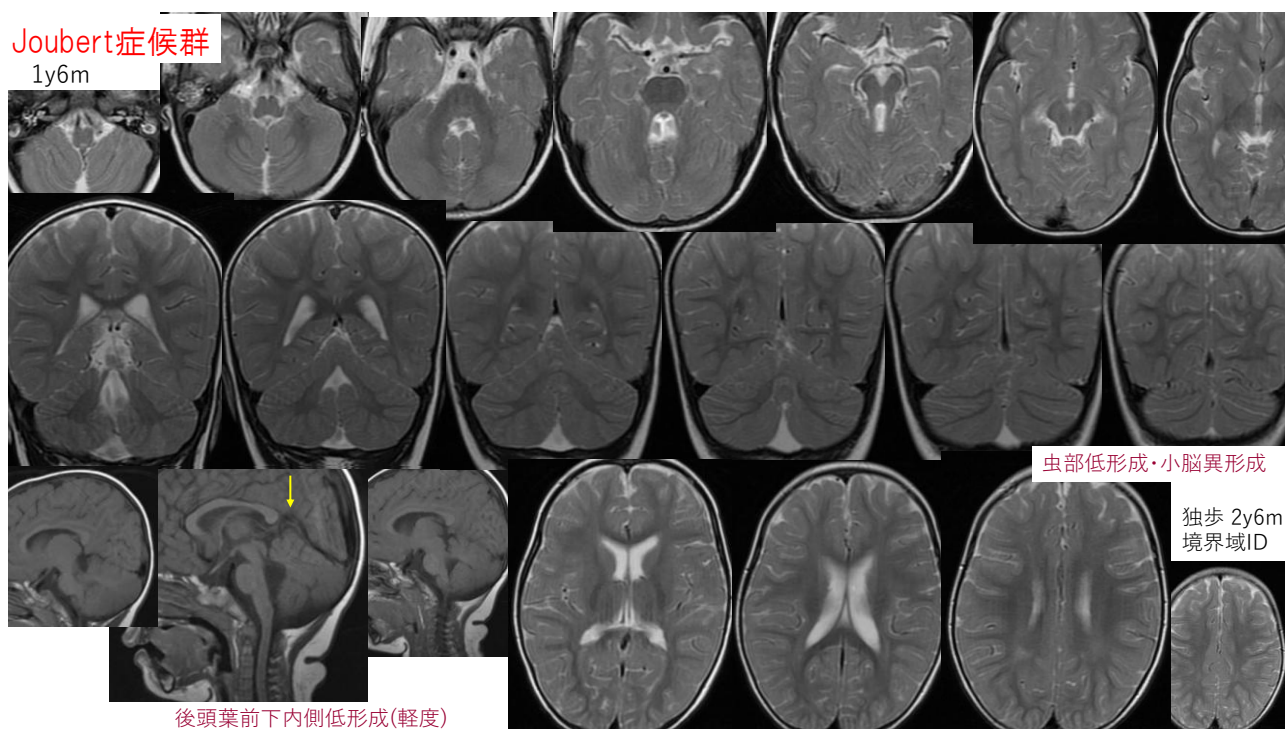
Joubert症候群



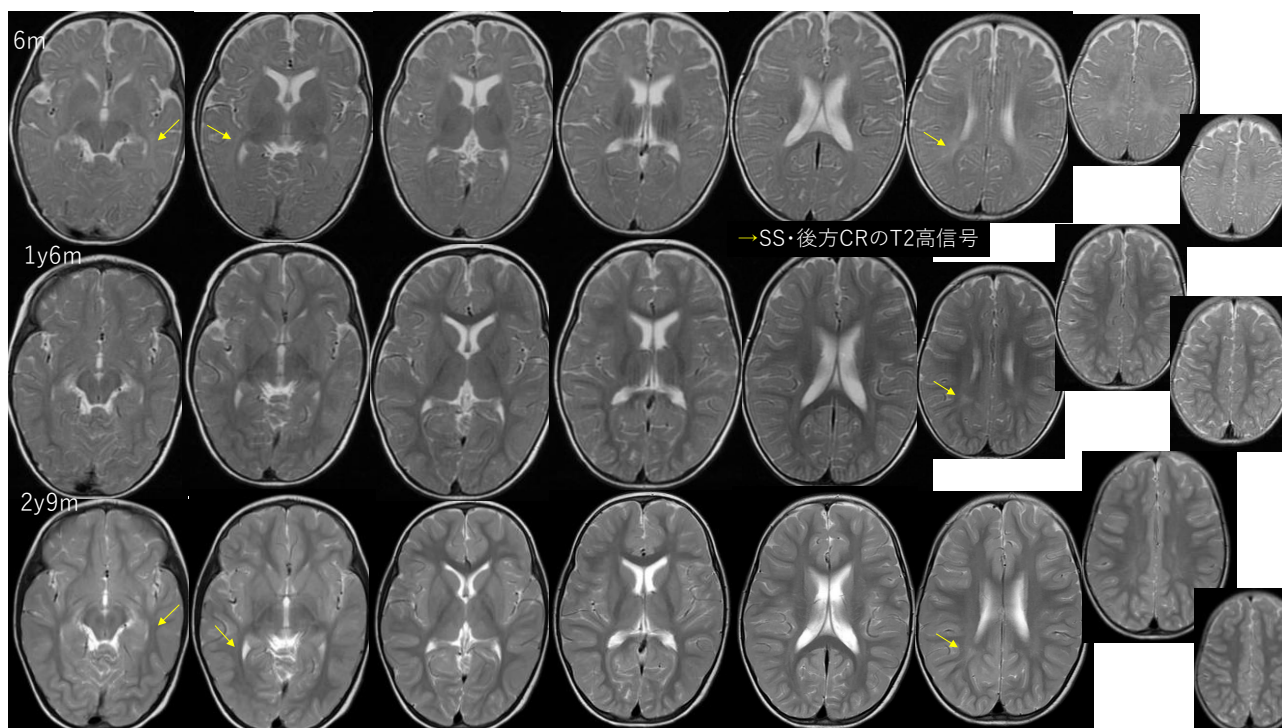
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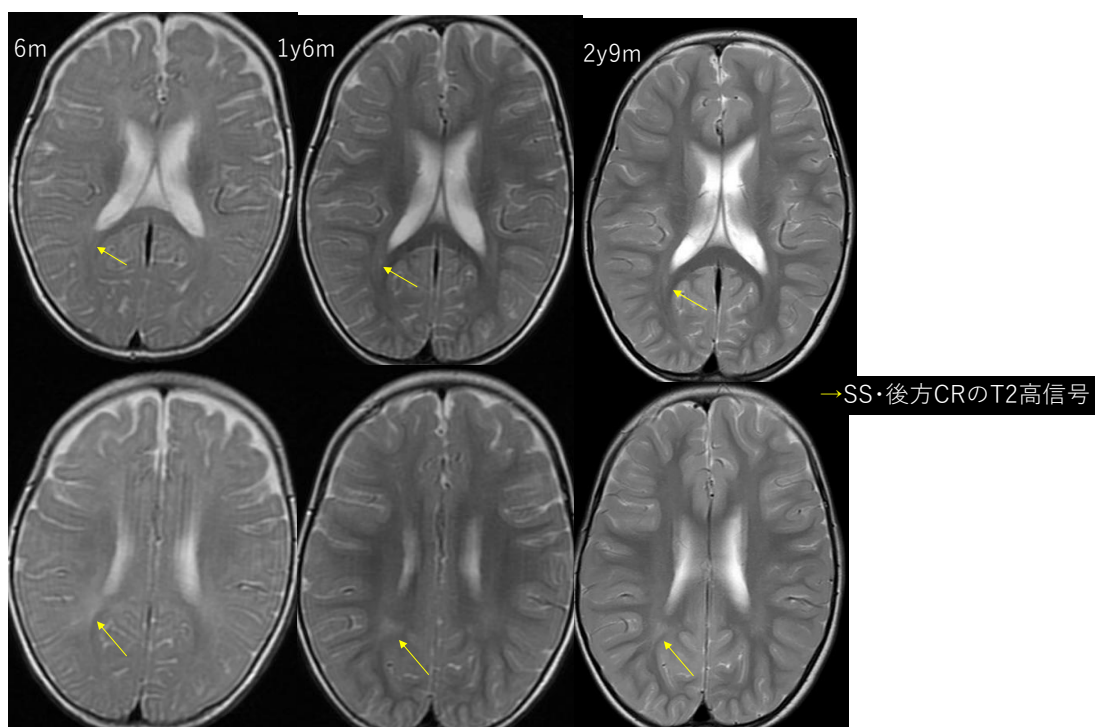
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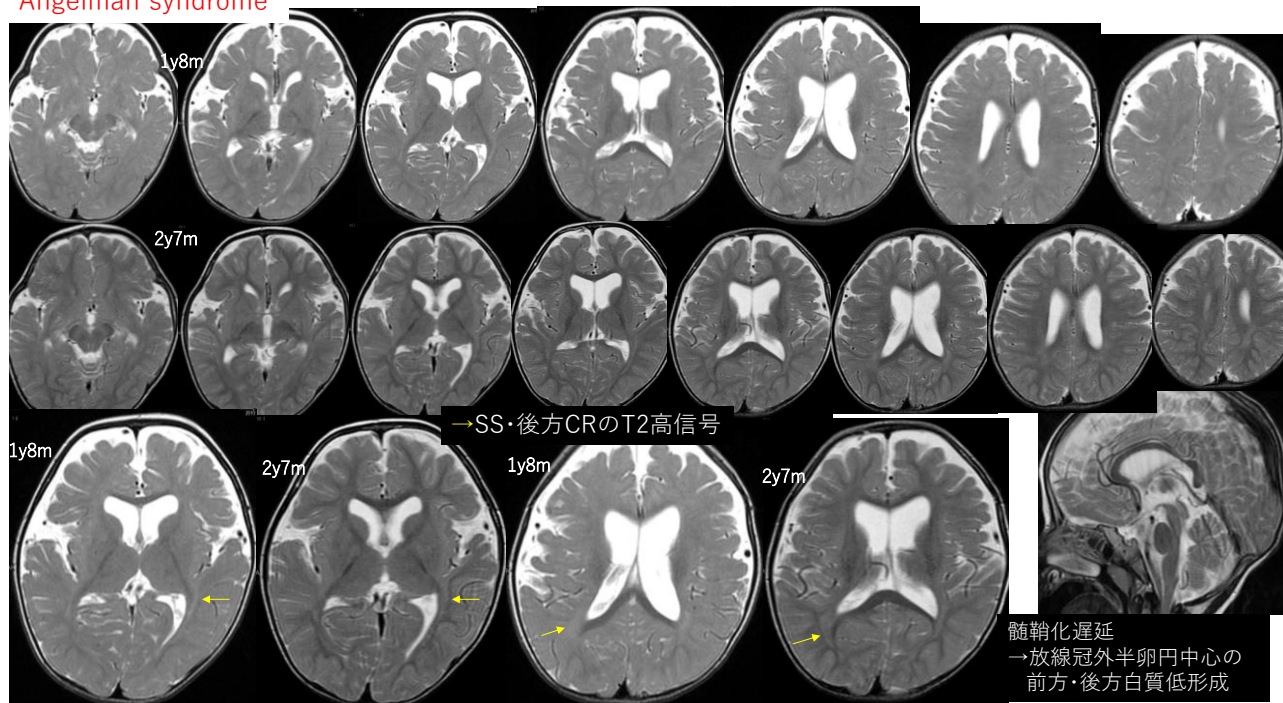


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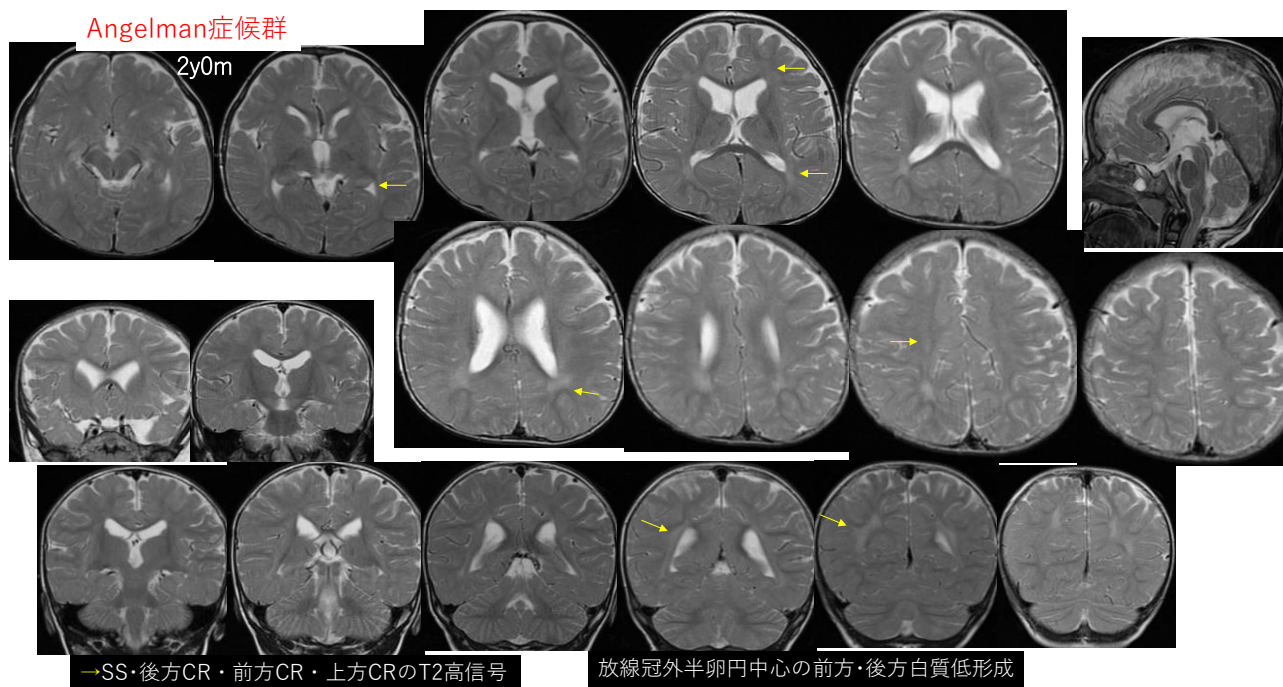
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Angelman syndrome



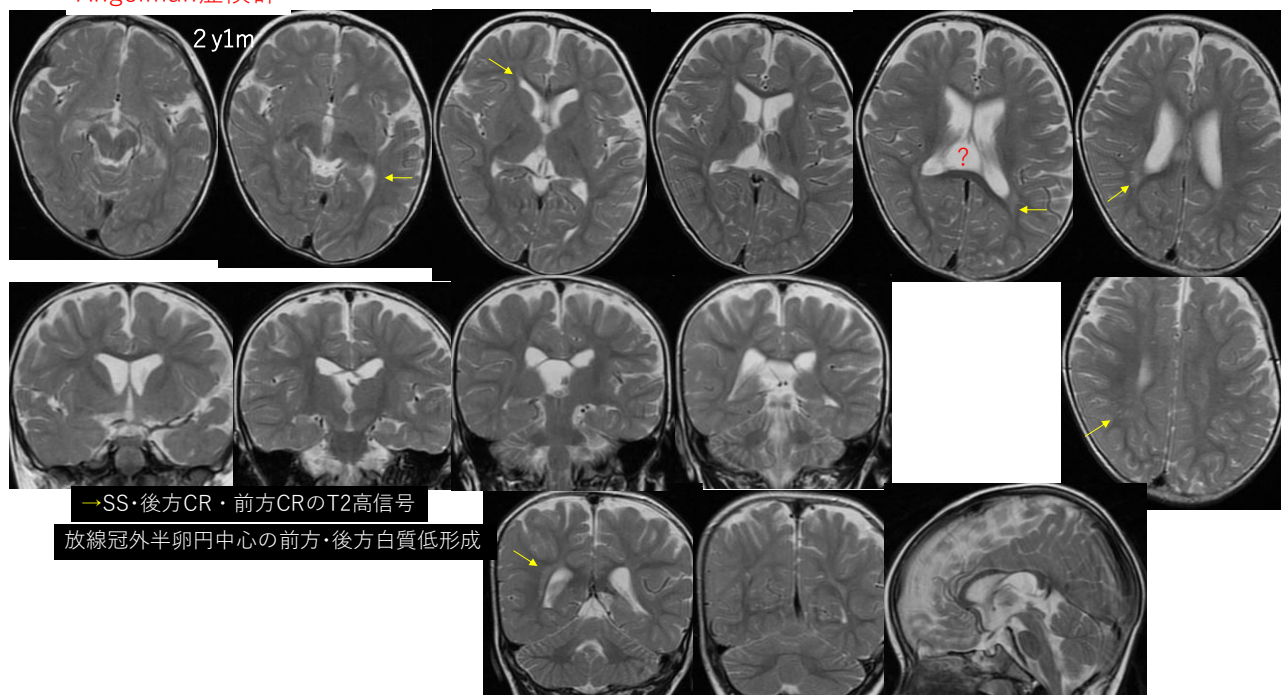
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Angelman症候群

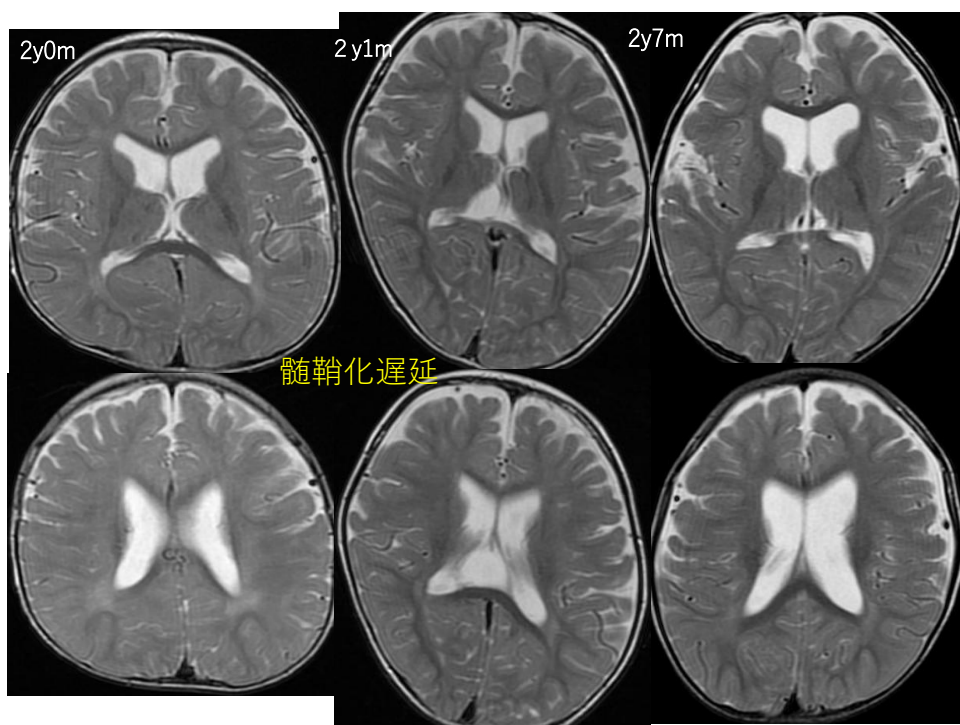


30

Angelman症候群

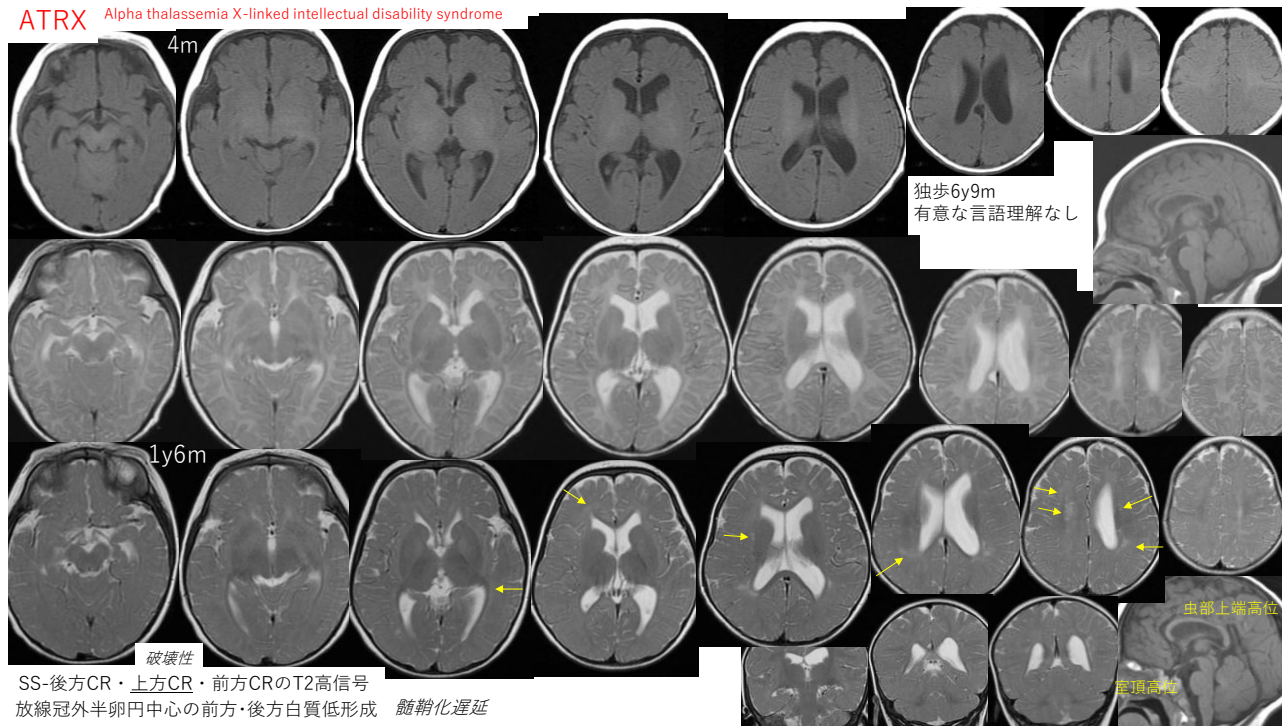


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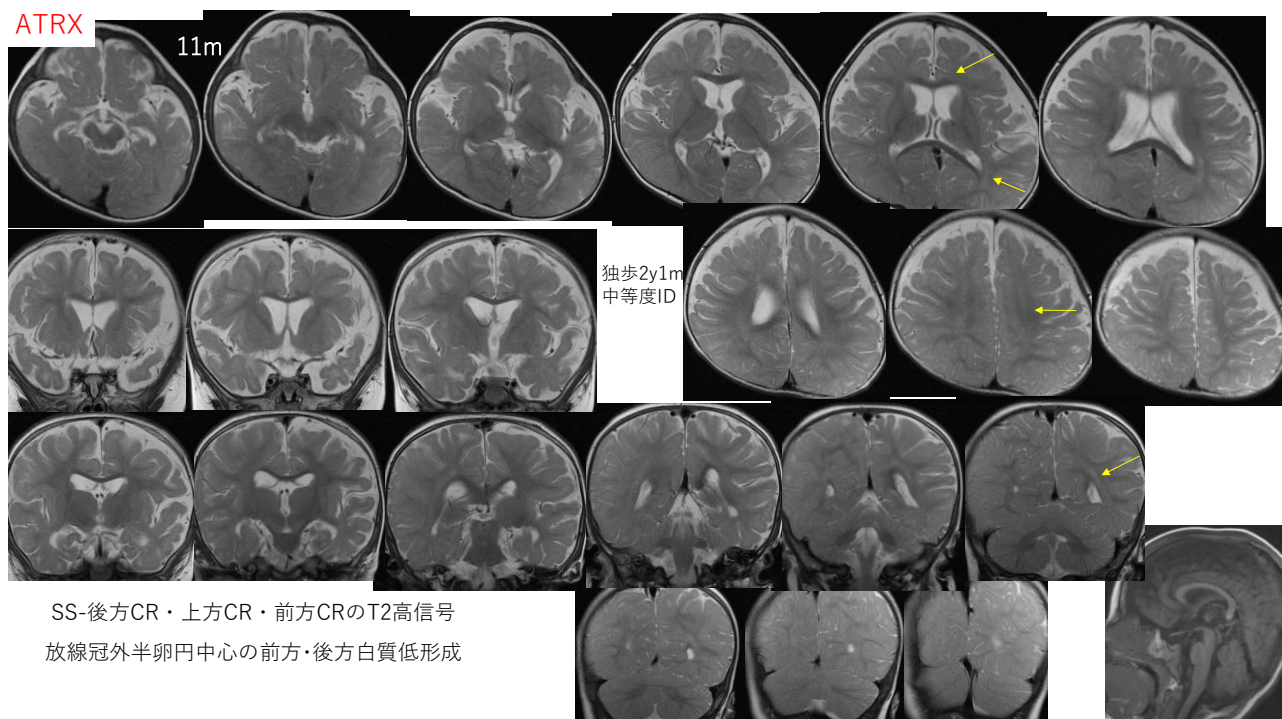
32

ATRX Alpha thalassemia X-linked intellectual disability syndrome



33

ATRX

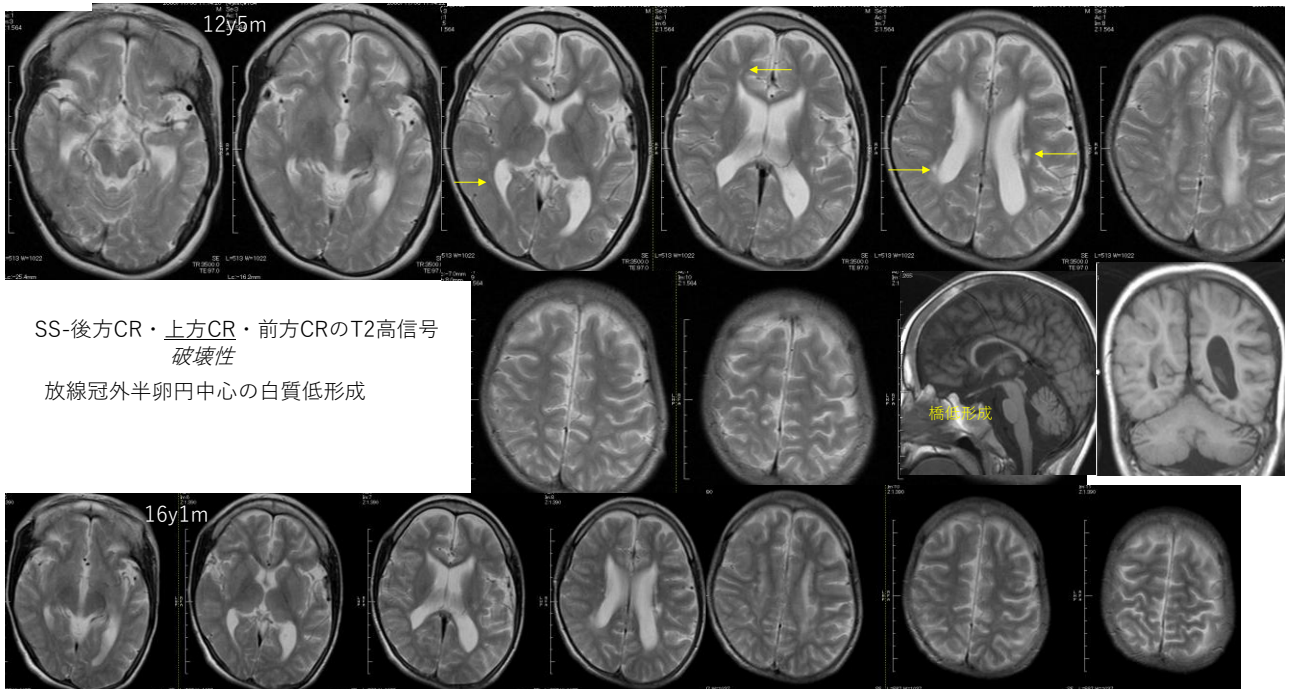


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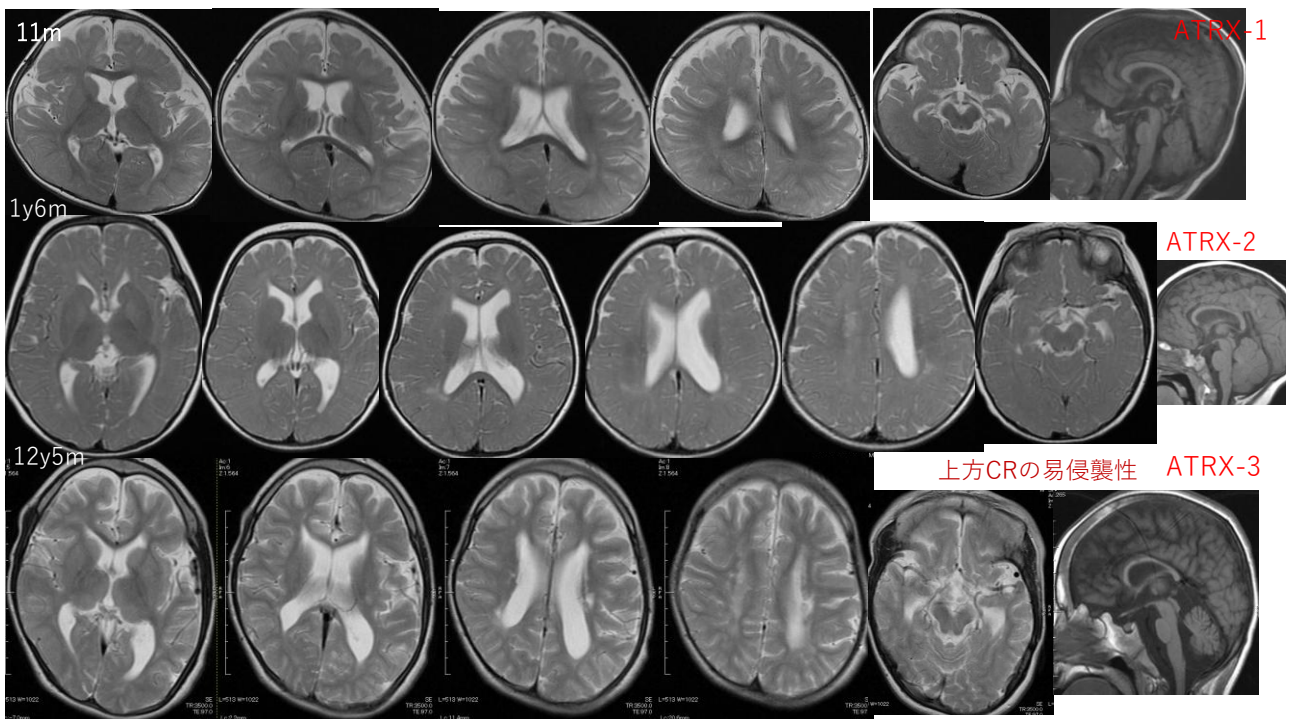
ATRX

3歳 室息

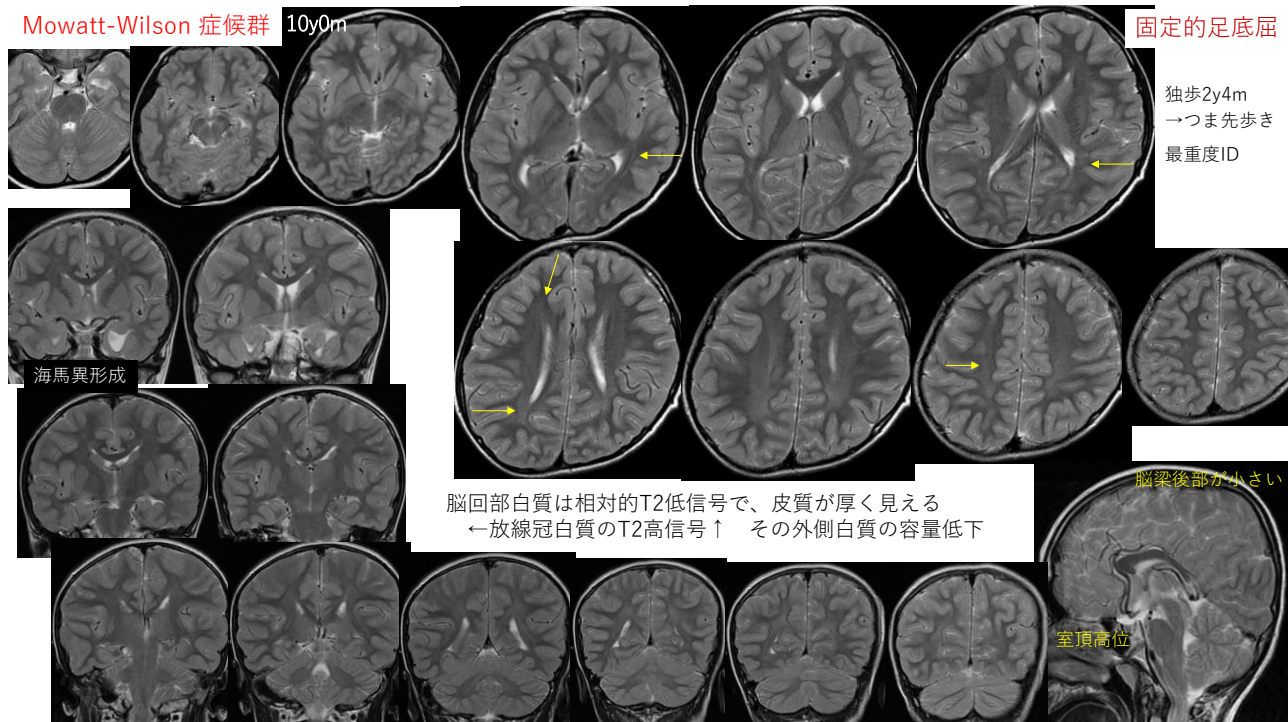
横地分類A1



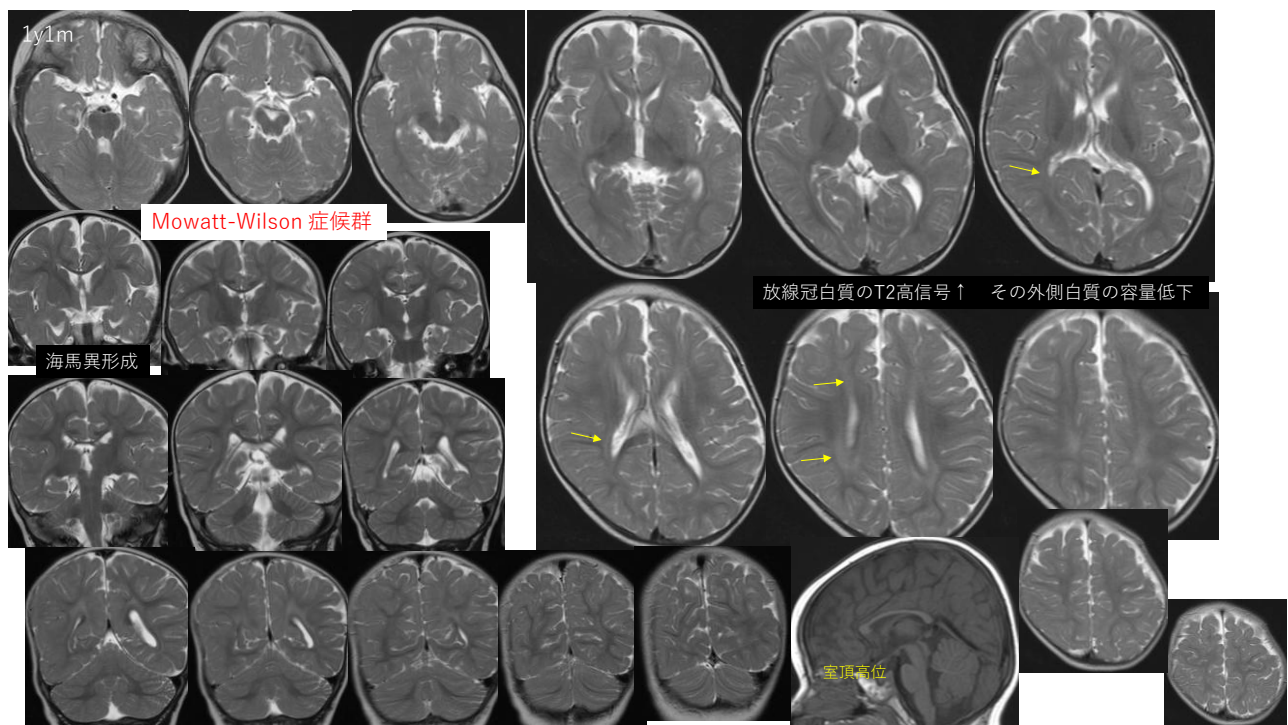
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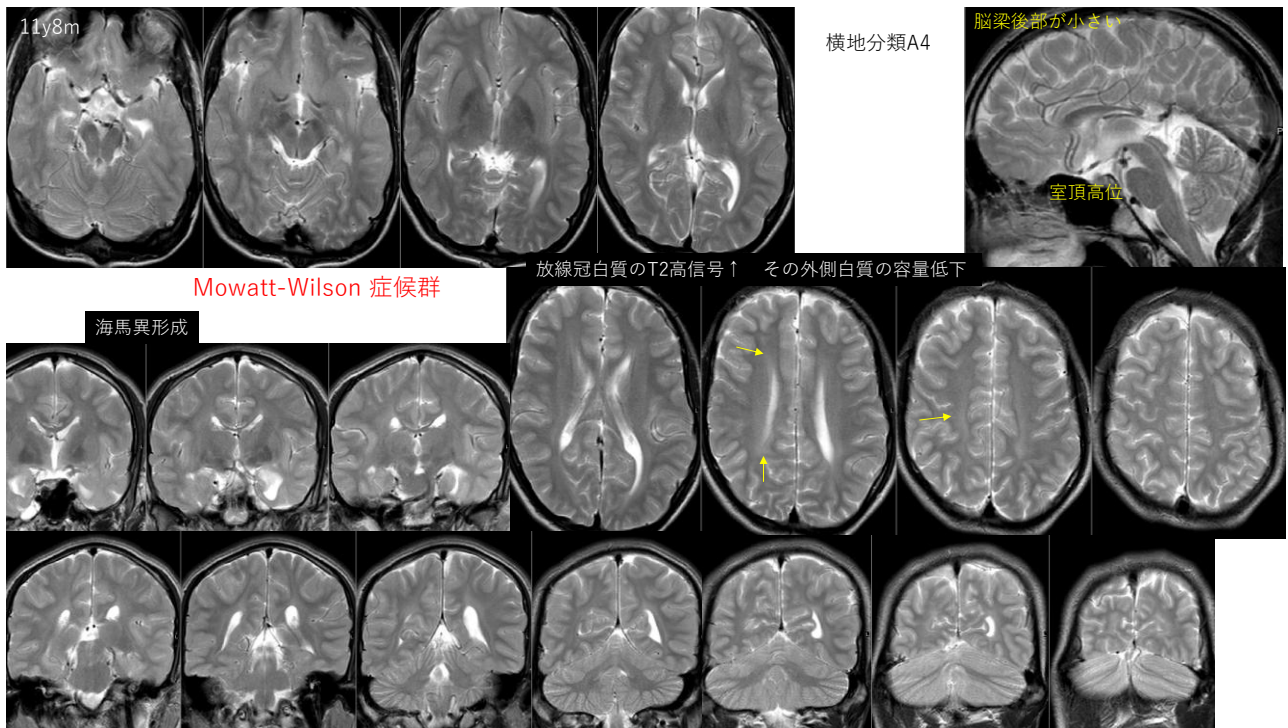
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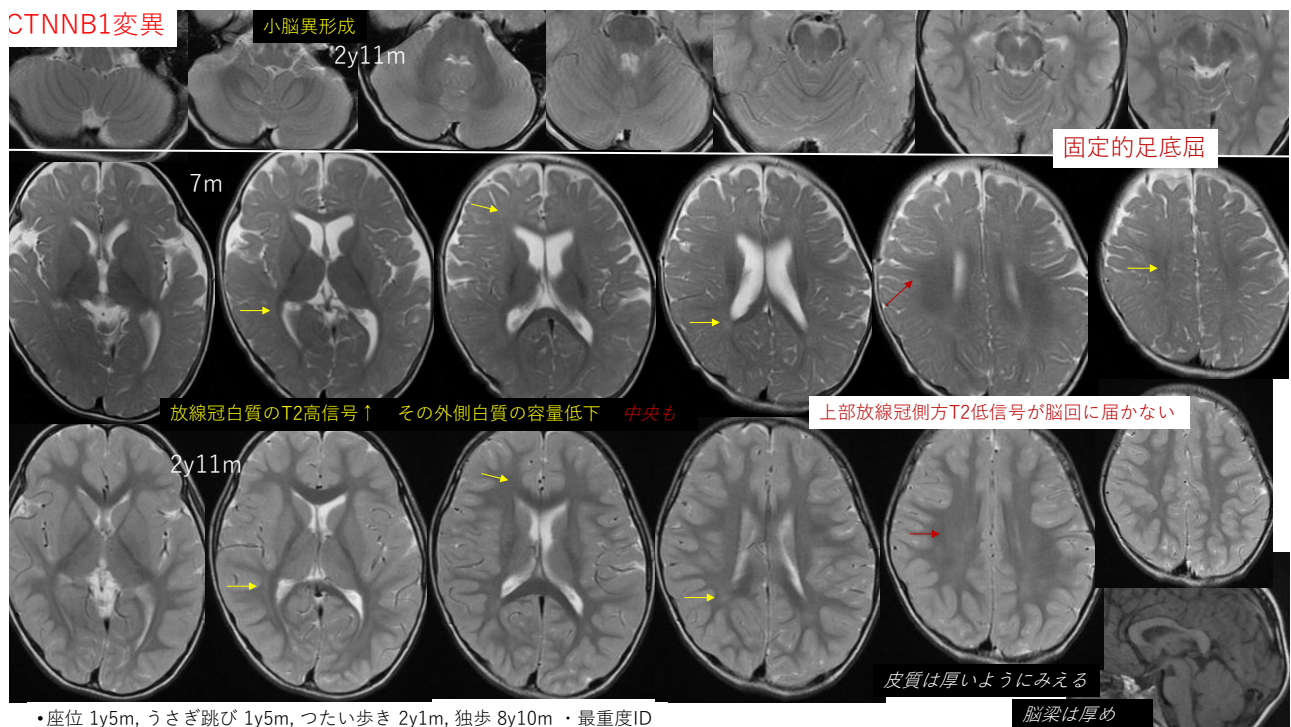
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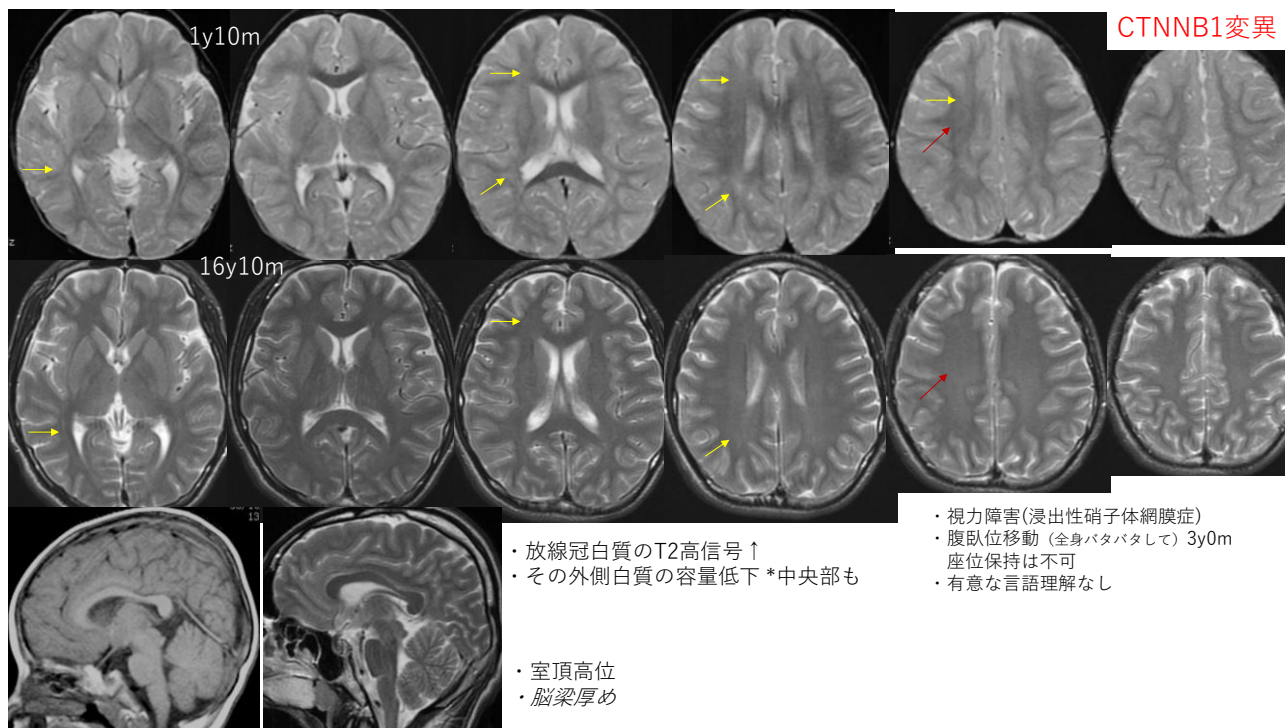
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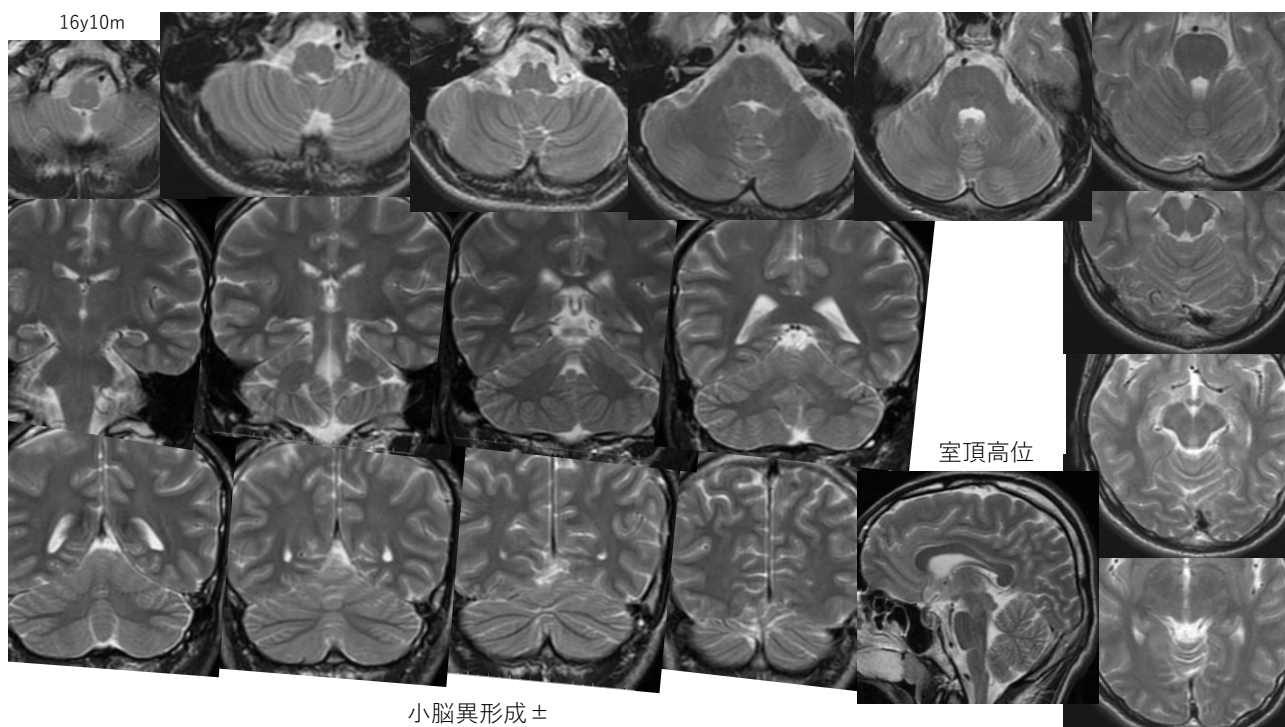
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40



41



42

