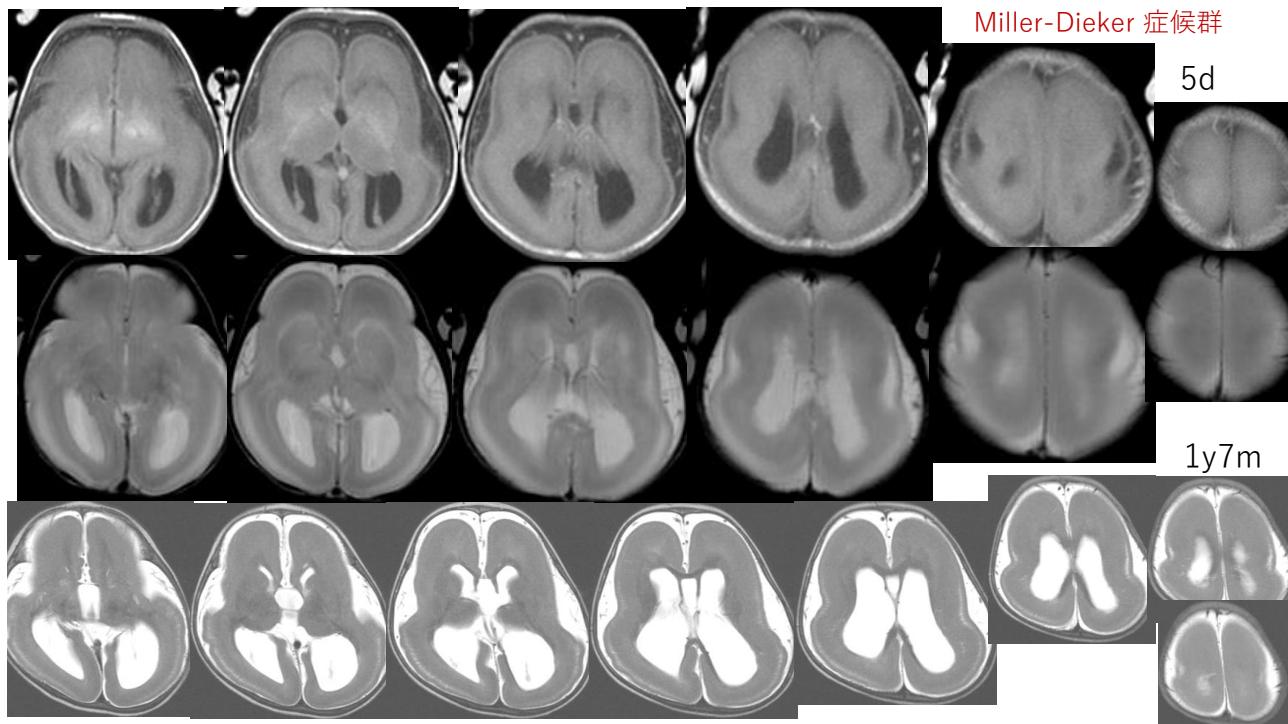


脳回形成異常の運動症候

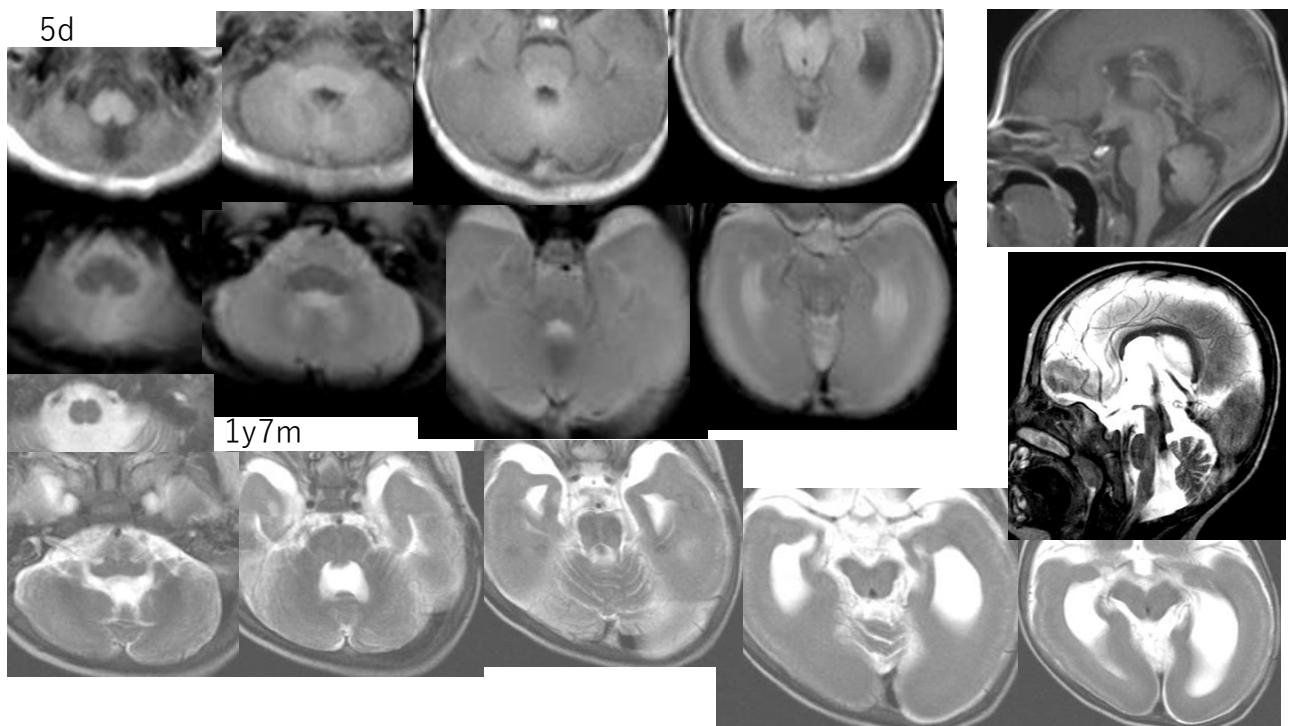
横地健治



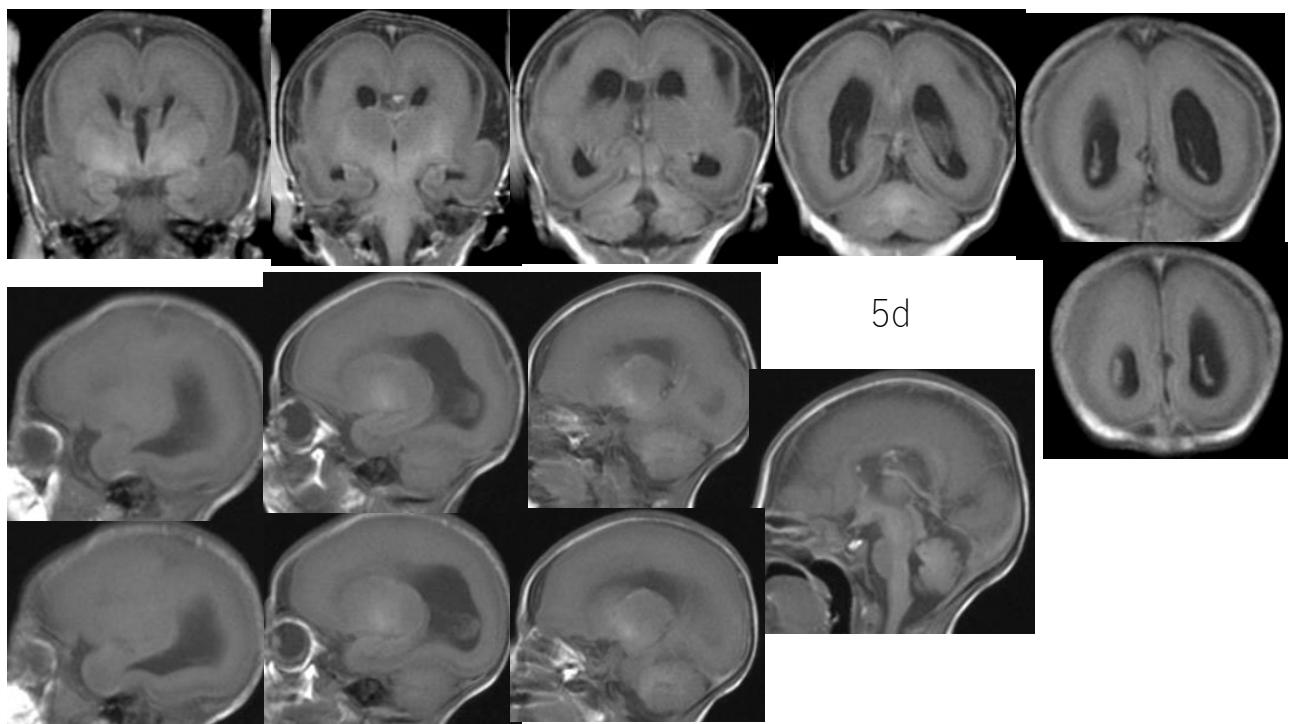
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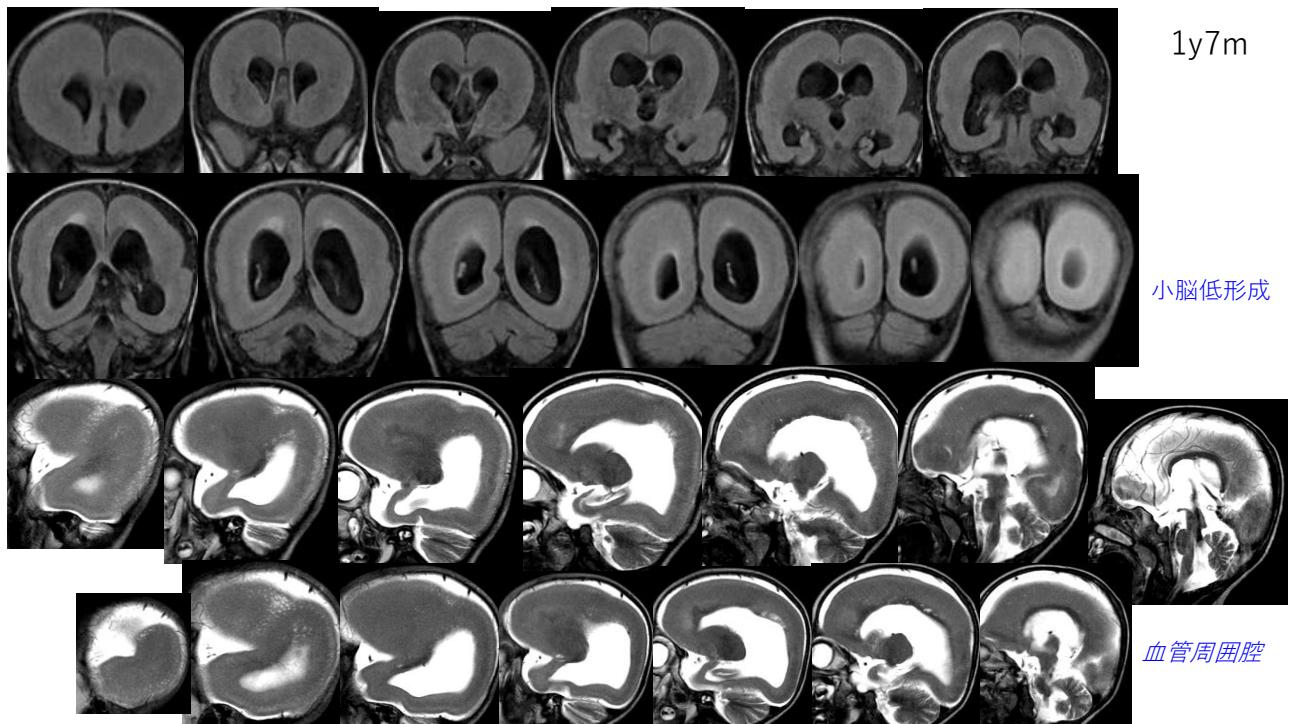
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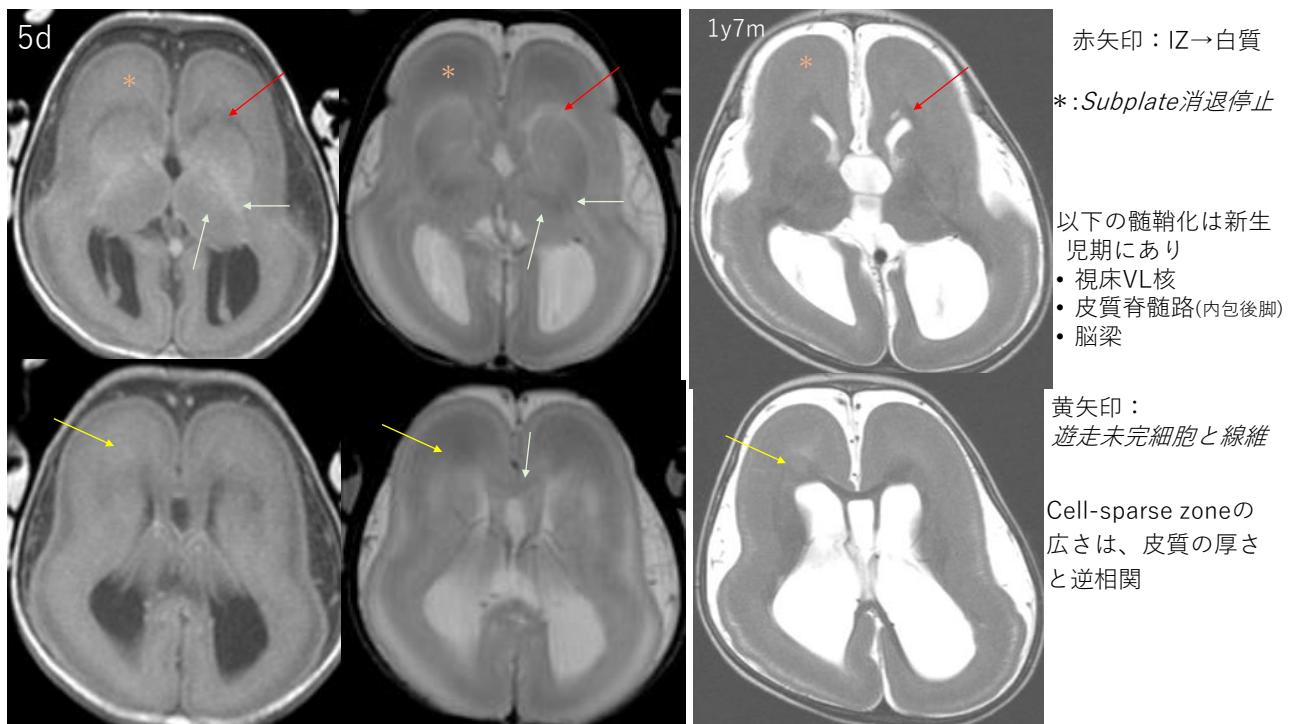
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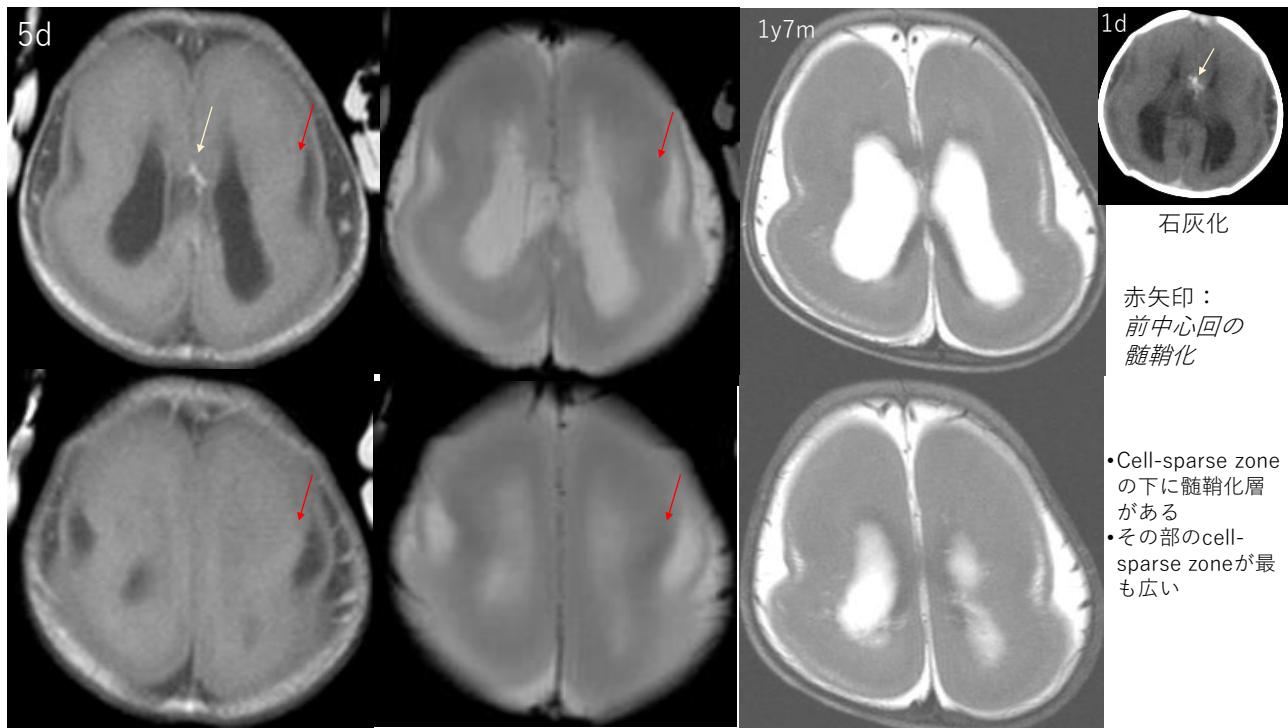
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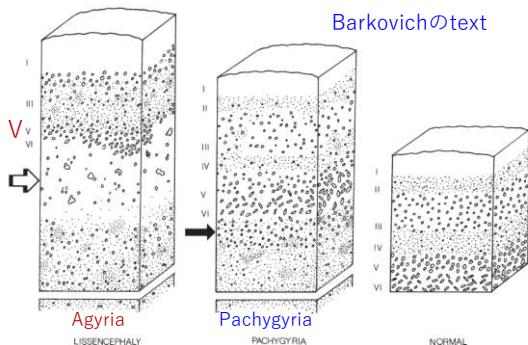
6



7

正常6層構造

- I : the molecular layer
- II : the external granular layer
- III: the external pyramidal layer
- IV: the internal granular layer
- V: the internal pyramidal layer
- VI: the multiform layer

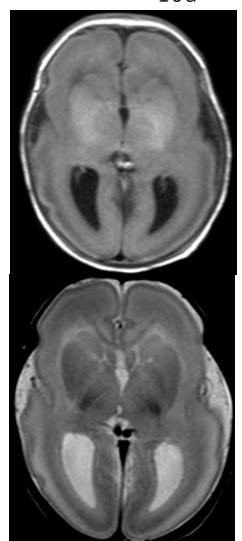


Schematic showing cortical architecture in classic lissencephaly. In complete **lissencephaly (agyria)**, a large cell-sparse zone (*open black arrow*) separates the molecular layer (layer I) and an outer cortical layer (layers III, V, VI in figure) from a thick deeper layer of disorganized neurons. In incomplete **lissencephaly (pachygryria)**, the outer cortical layer is thicker, the cell-sparse zone (*black arrow*) thinner, and the inner cortical layer smaller. **?**

Forman MS, Squier W, Dobyns WB, Golden JA. Genotypically defined lissencephalies show distinct pathologies. *J Neuropathol Exp Neurol*. 2005;64:847-57.

8

Miller-Dieker 症候群



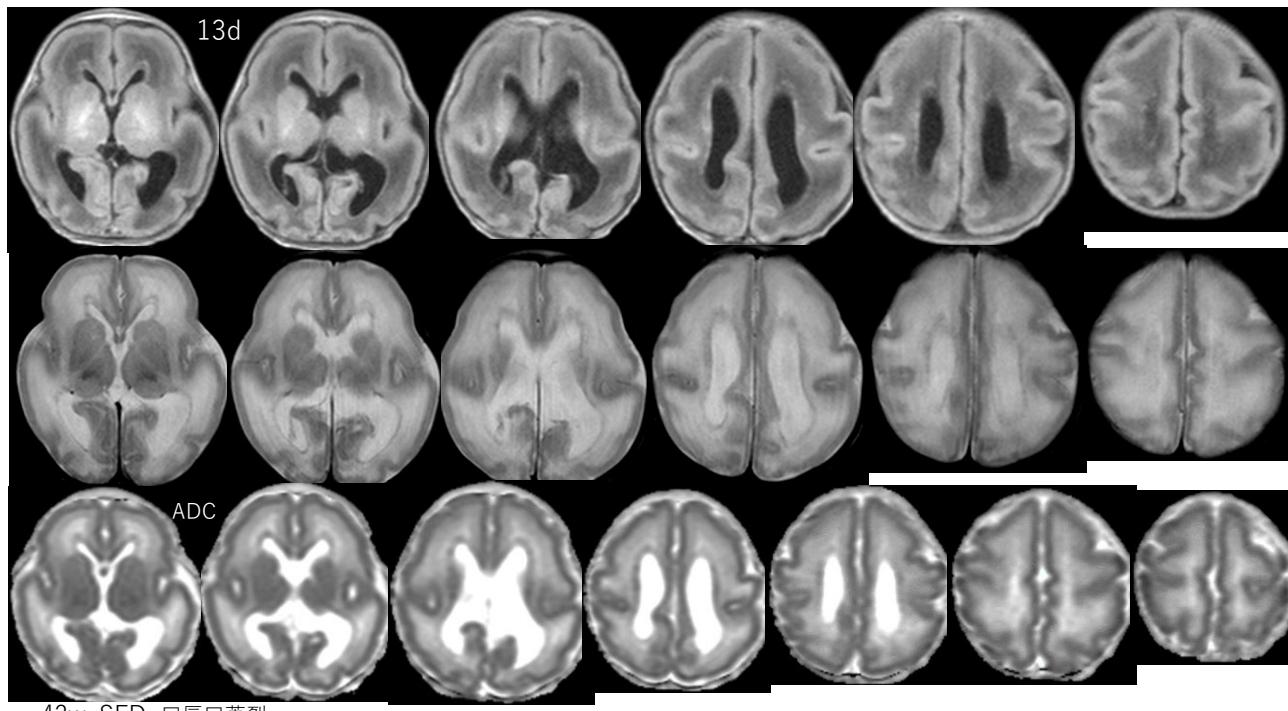
股屈曲過活動 股伸展荷重制限 共収縮制御障害

- ・頸部伸展・体幹伸展は変わらず
- ・左股伸展は緩み、膝は屈曲する 自然経過

9

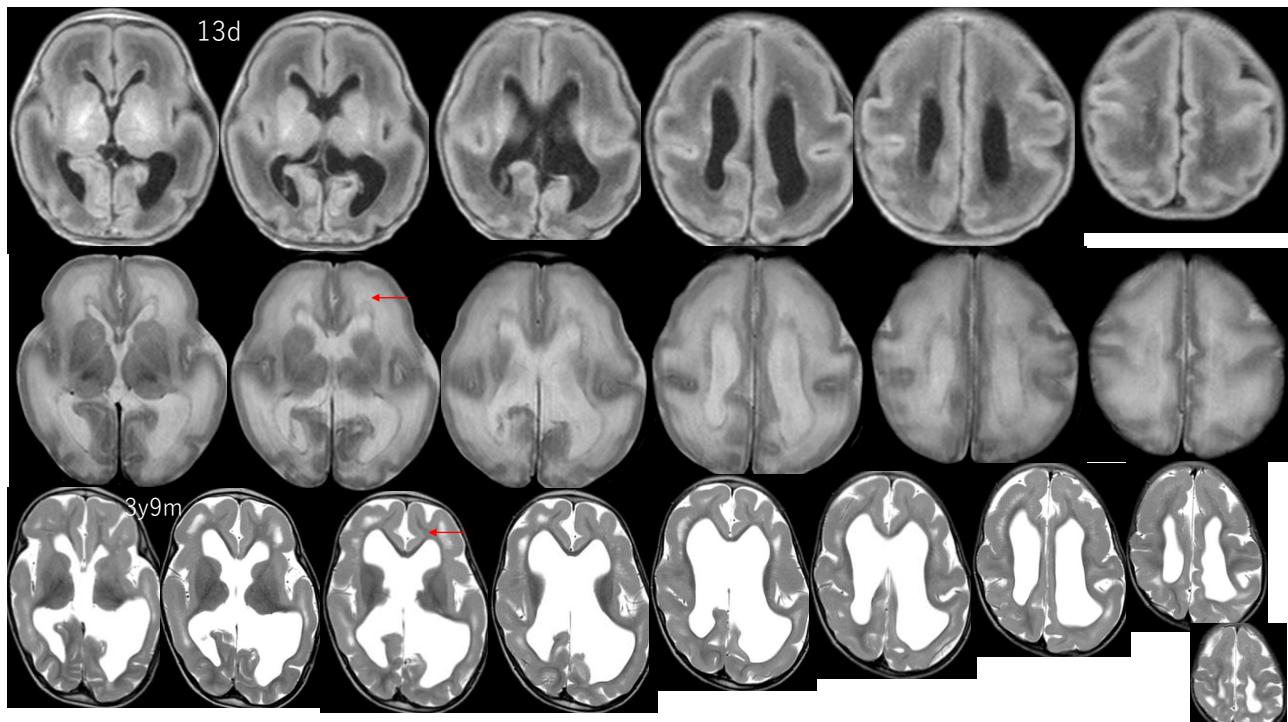


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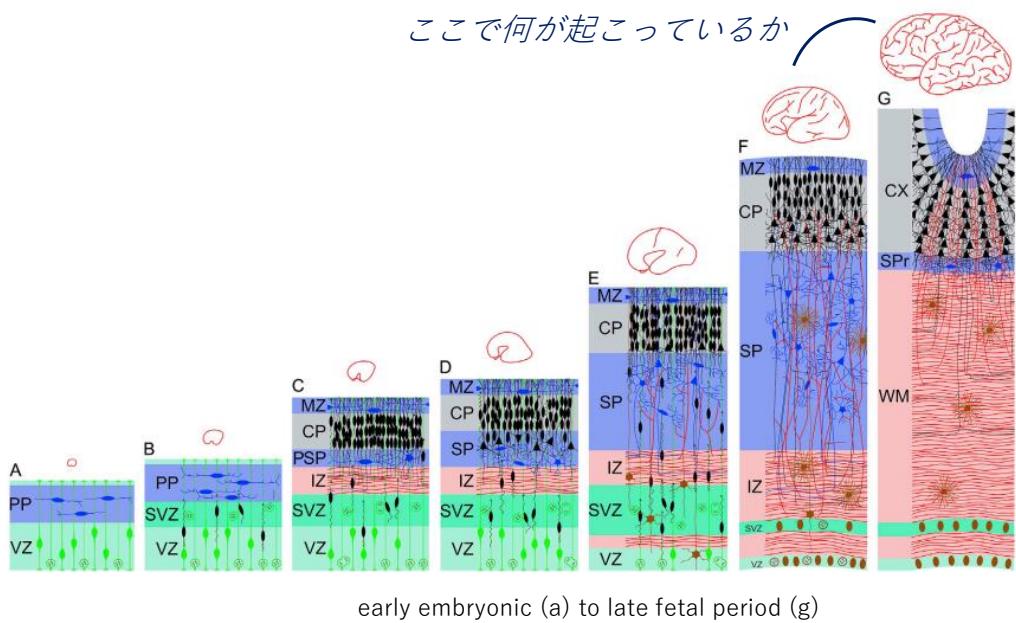


42w, SFD 口唇口蓋裂

11



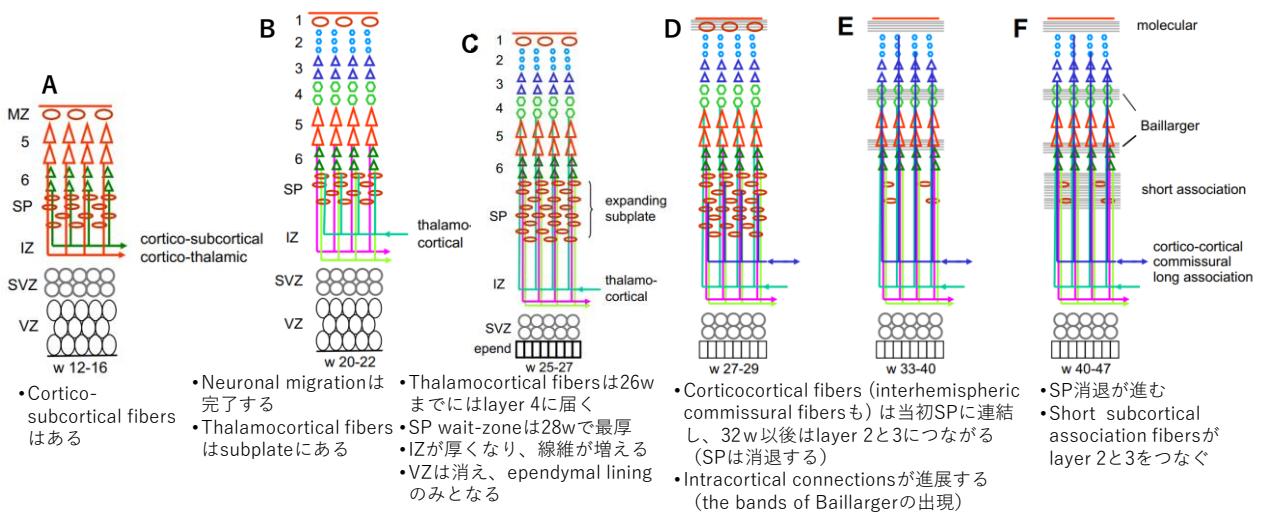
12



Kostovic I, et al. Neural histology and neurogenesis of the human fetal and infant brain. *NeuroImage* 2019;188:743-73.

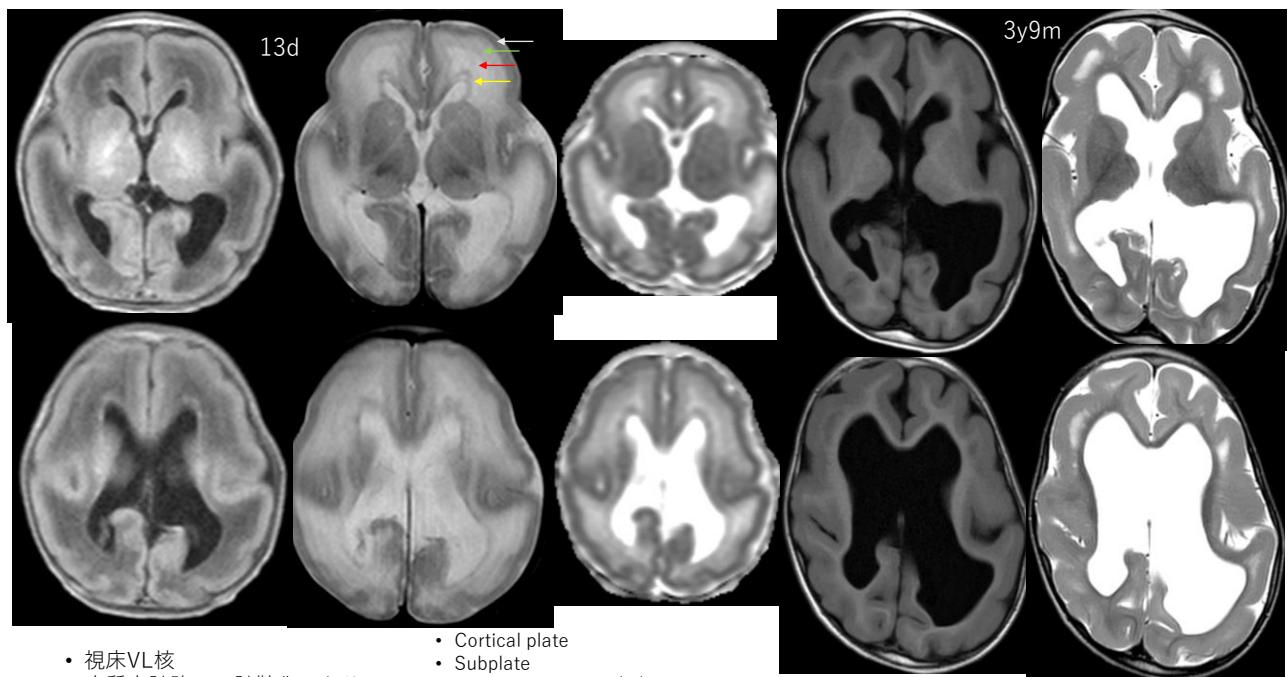
13

Development of the connectivity



Raybaud C, Widjaja E. Development and dysgenesis of the cerebral cortex: malformations of cortical development. *Neuroimaging Clin N Am* 2011;21:483-543.

14

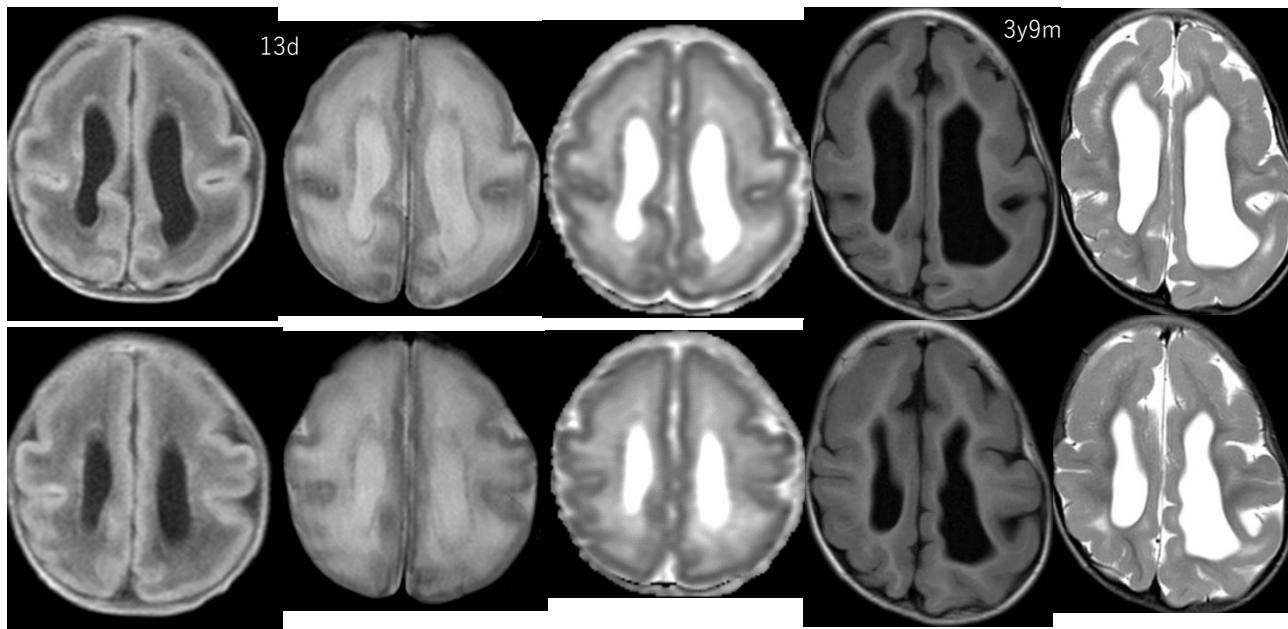


- 視床VL核
- 皮質脊髄路 の髓鞘化はあり

- Cortical plate
- Subplate
- Intermediate zone (IZ)
- Subventricular zone ~ ventricular zone でいいが

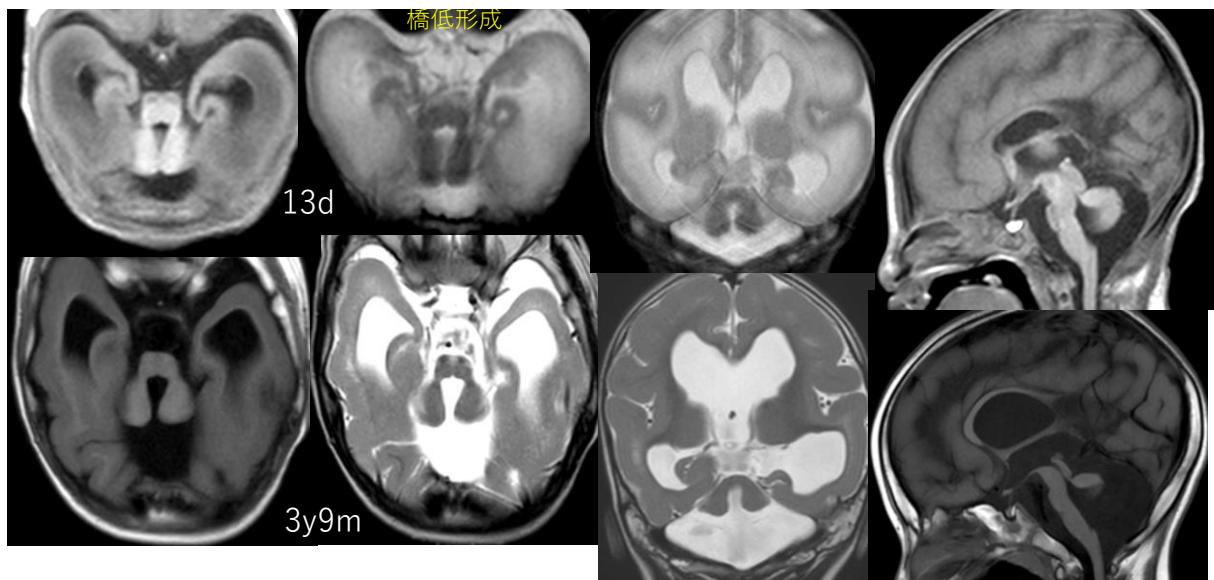
脳溝は深くはなっているが、数は増えず

15



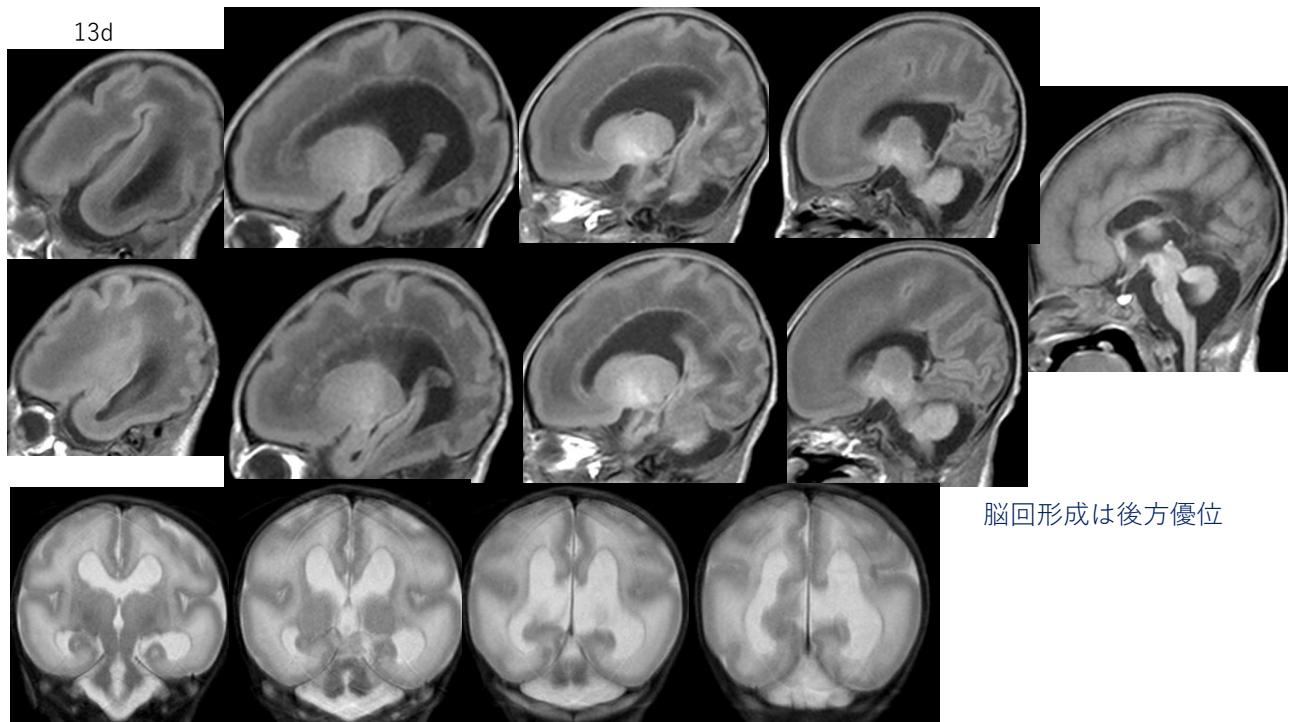
中心溝あたりの脳回は髓鞘化あり

16

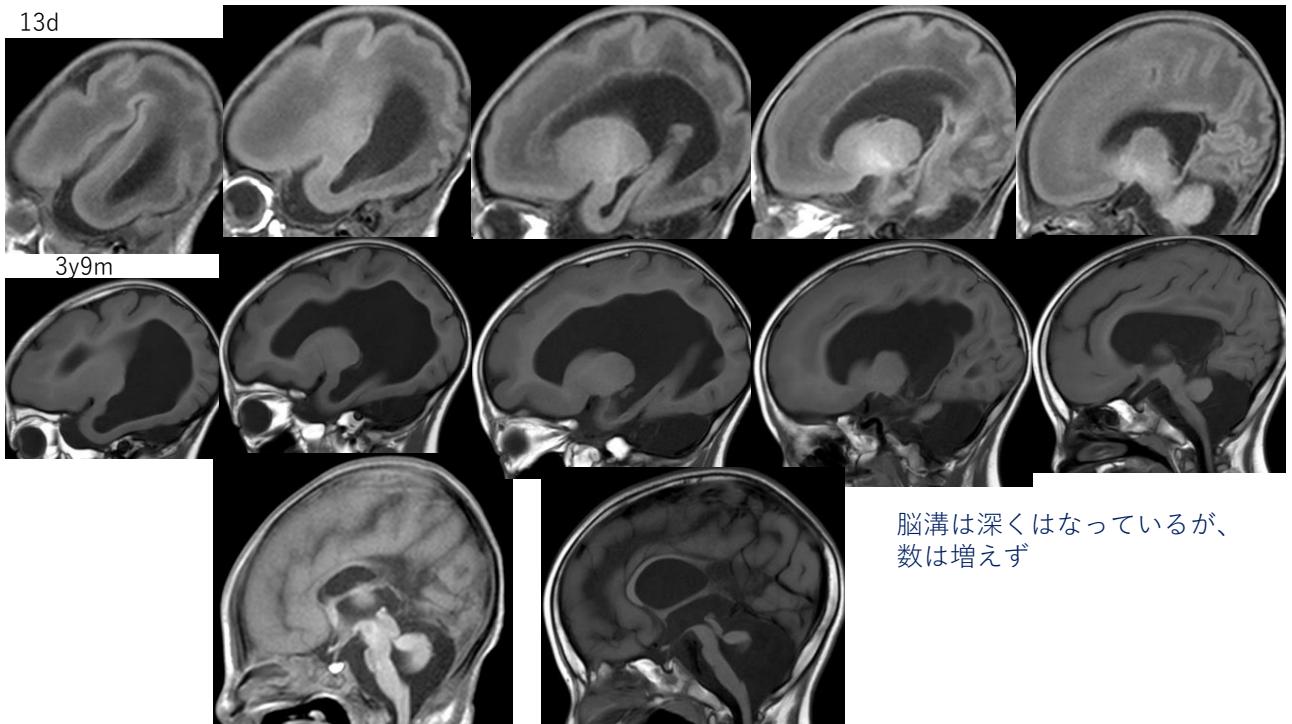


- 中小脳脚→小脳半球 系の無形成
- 虫部無形成
- ✓ 下小脳脚系の過形成

17



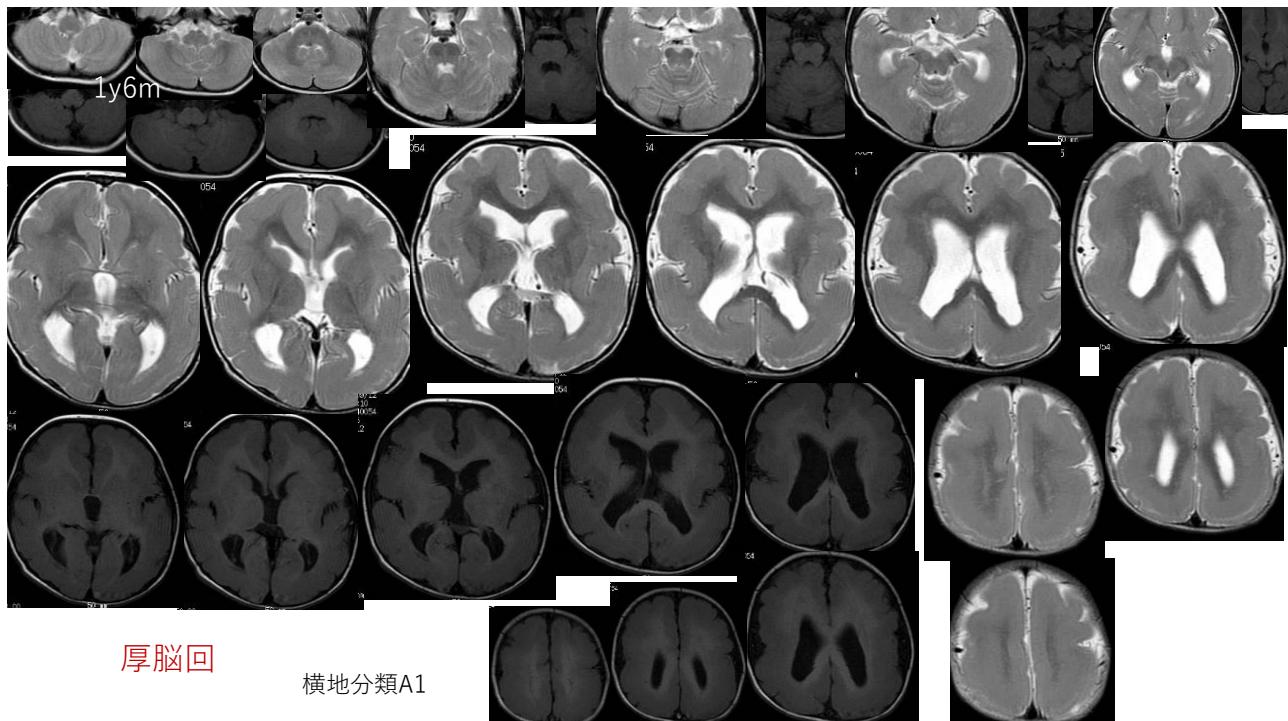
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19



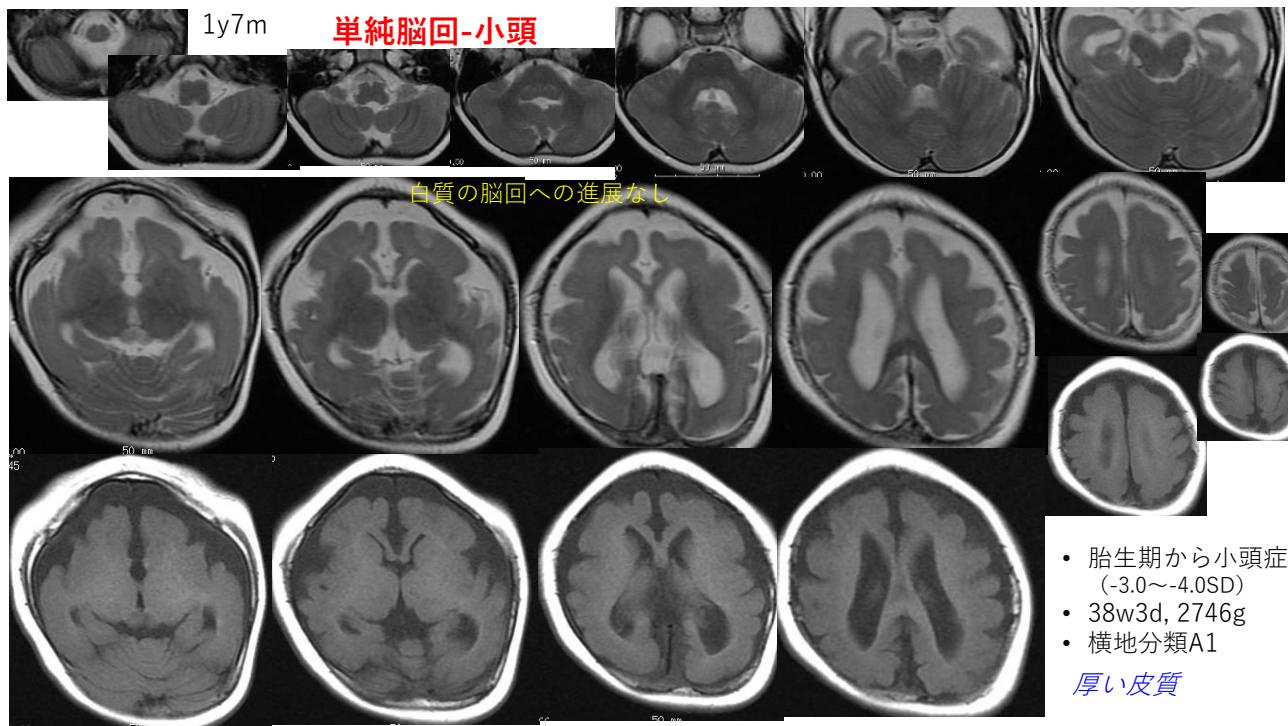
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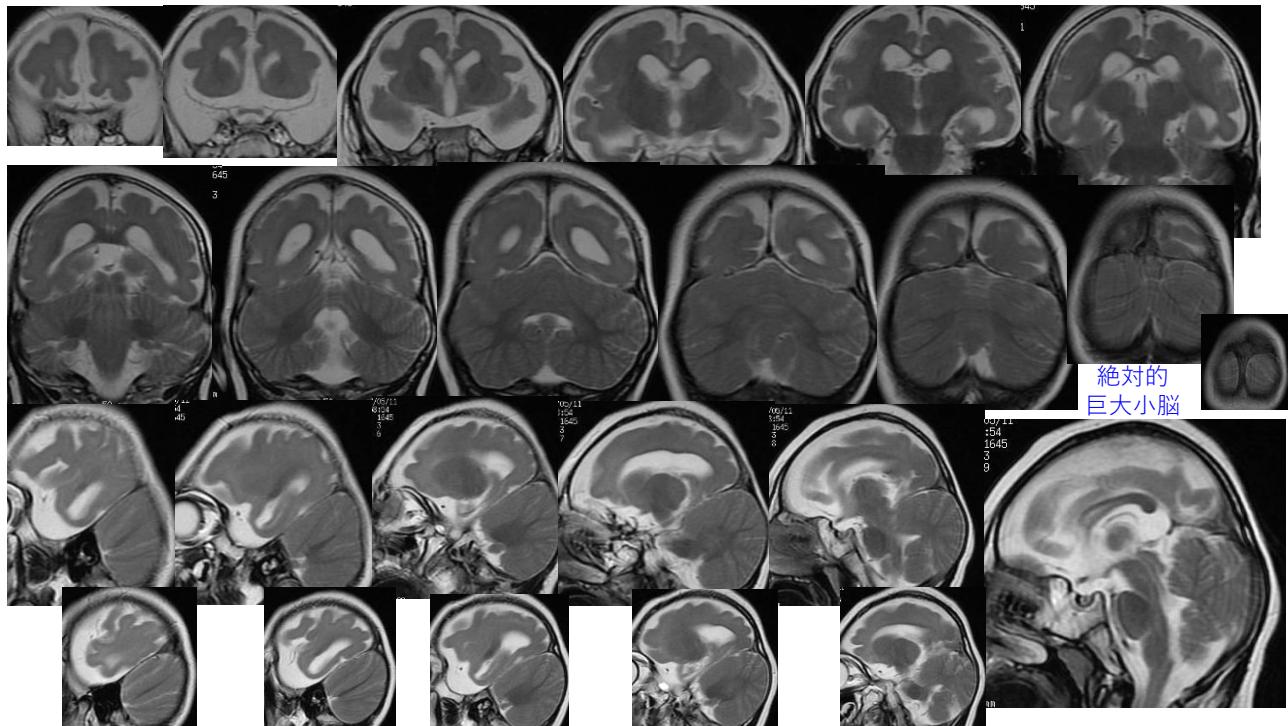
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22



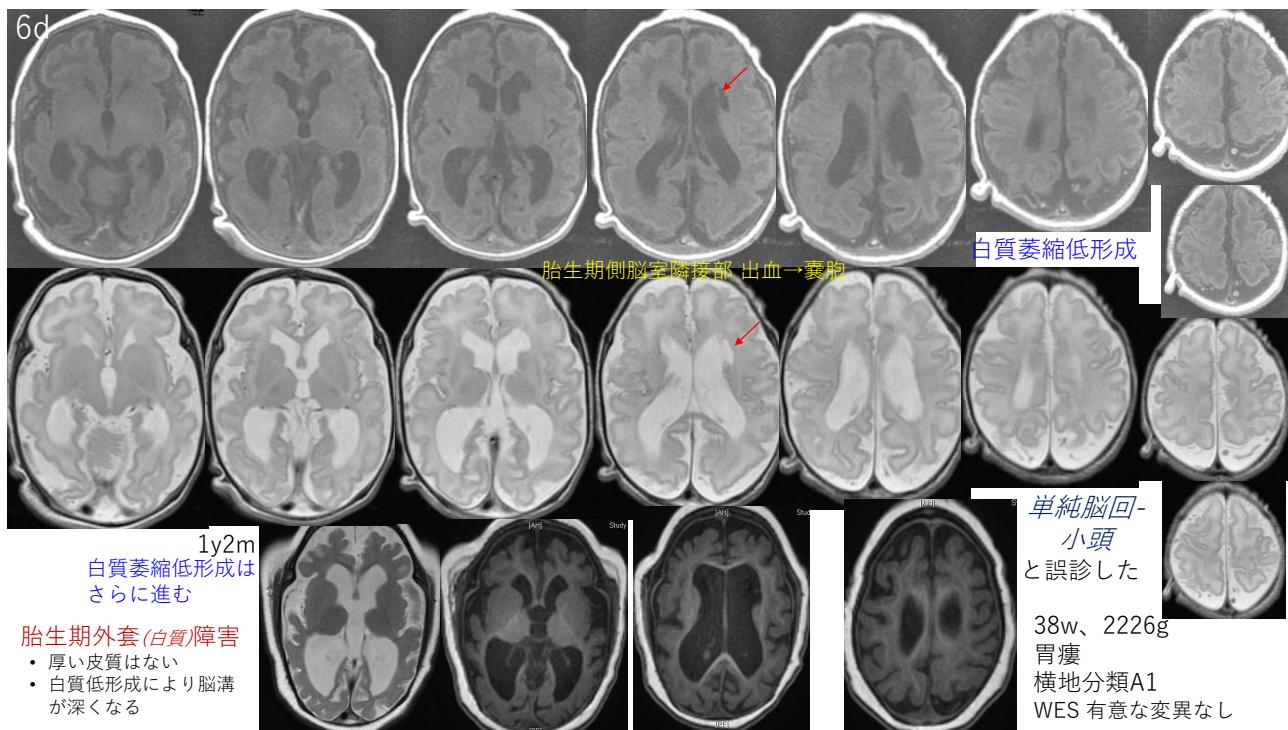
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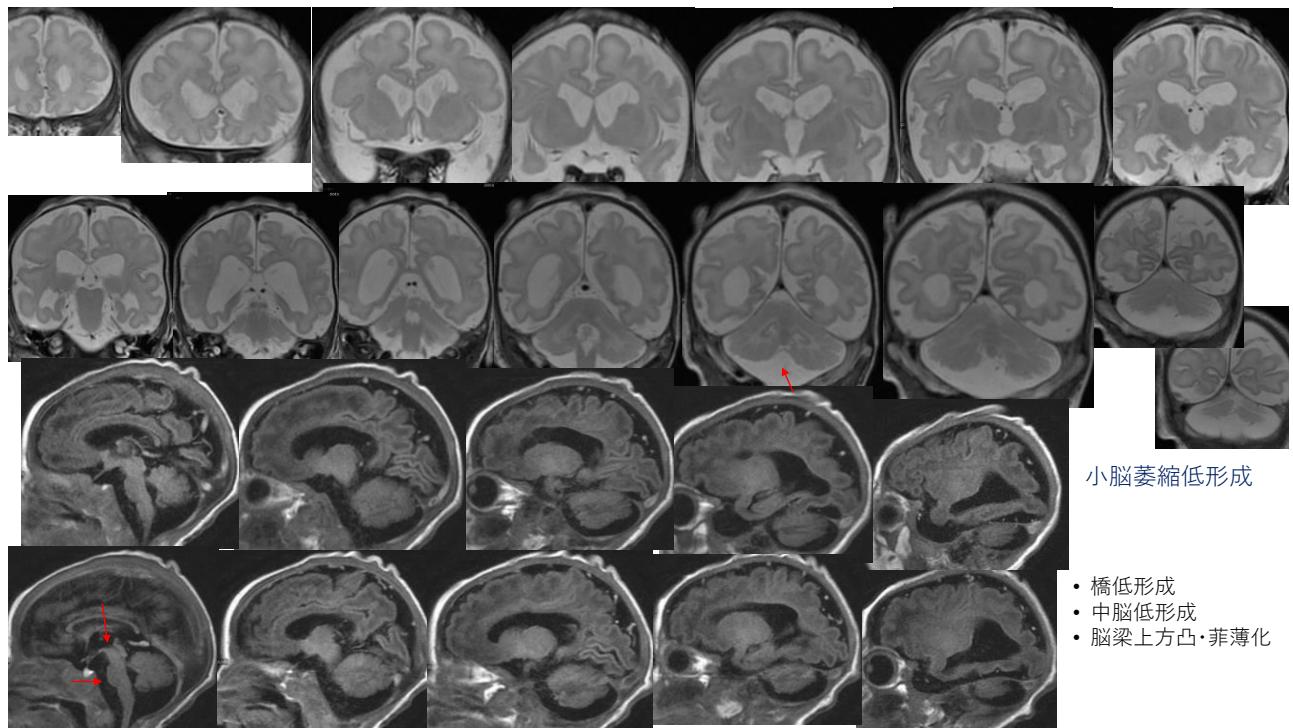
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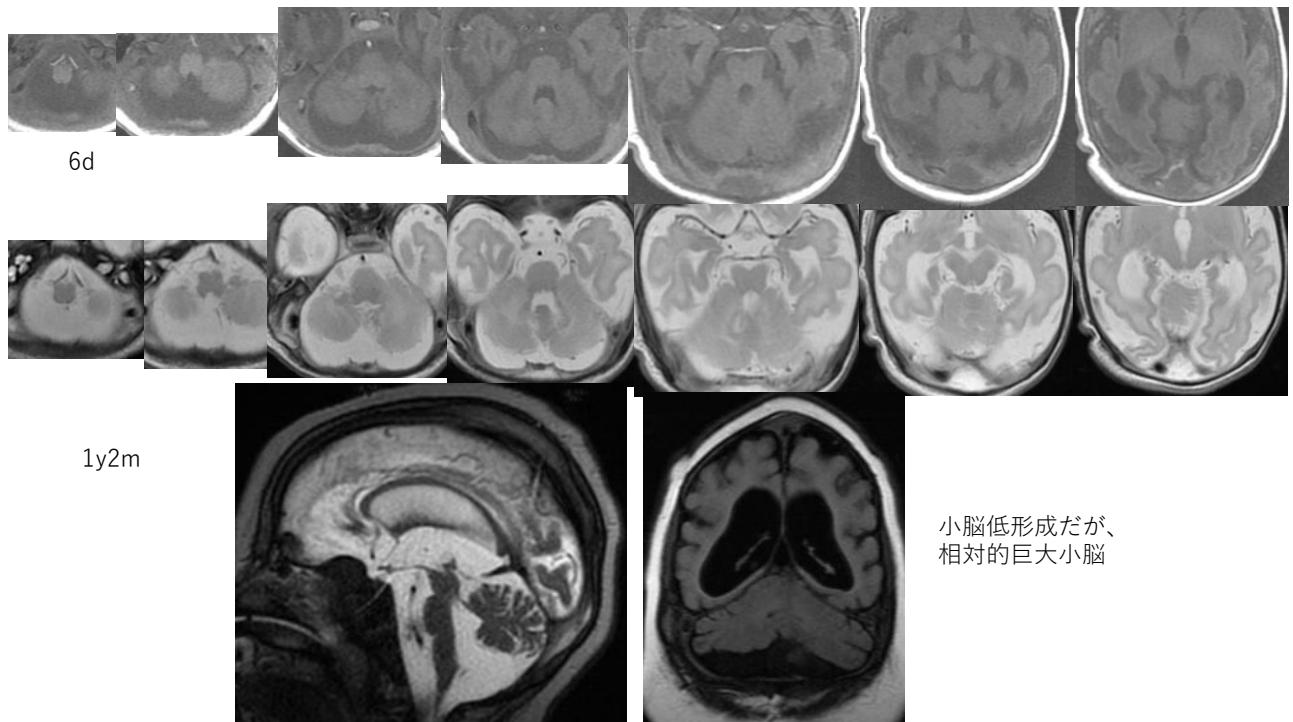
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26



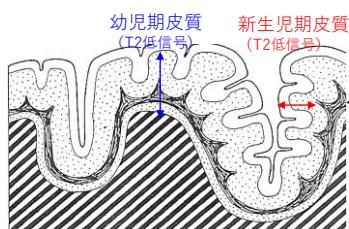
27



28



29



Polymicrogyria (PMG)

4層構造の第3層は、新生児期T2高信号で、その後髓鞘化しT2低信号となる。
 Takanashi J, Barkovich AJ. The changing MR imaging appearance of polymicrogyria: a consequence of myelination. AJNR 2003;24:788-93.

Jansen AC, et al. **The histopathology of polymicrogyria**: a series of 71 brain autopsy studies. Dev Med Child Neurol 2016;58:39-48.

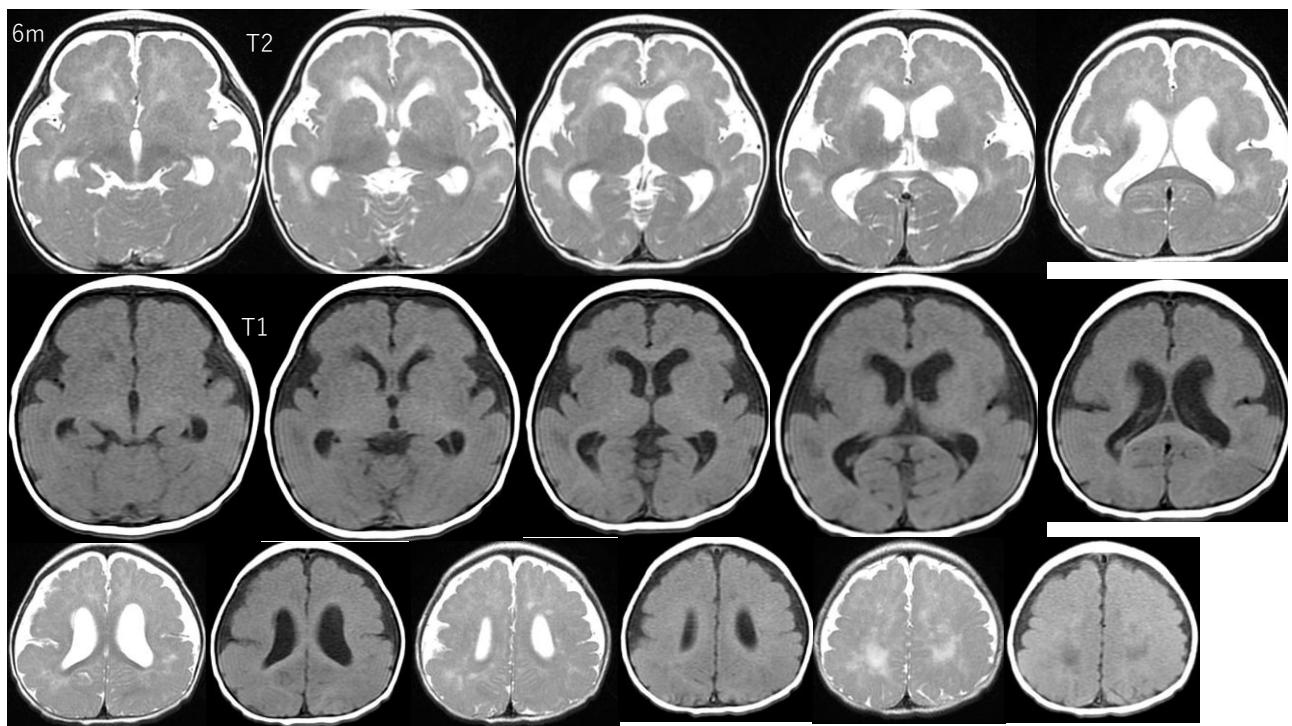
NormanのPMG分類

- Unlayered cortex (*Two-layer*)
- **Four-layered cortex**
- Parallel four-layered cortex
- Miniature gyri
- Poorly laminated

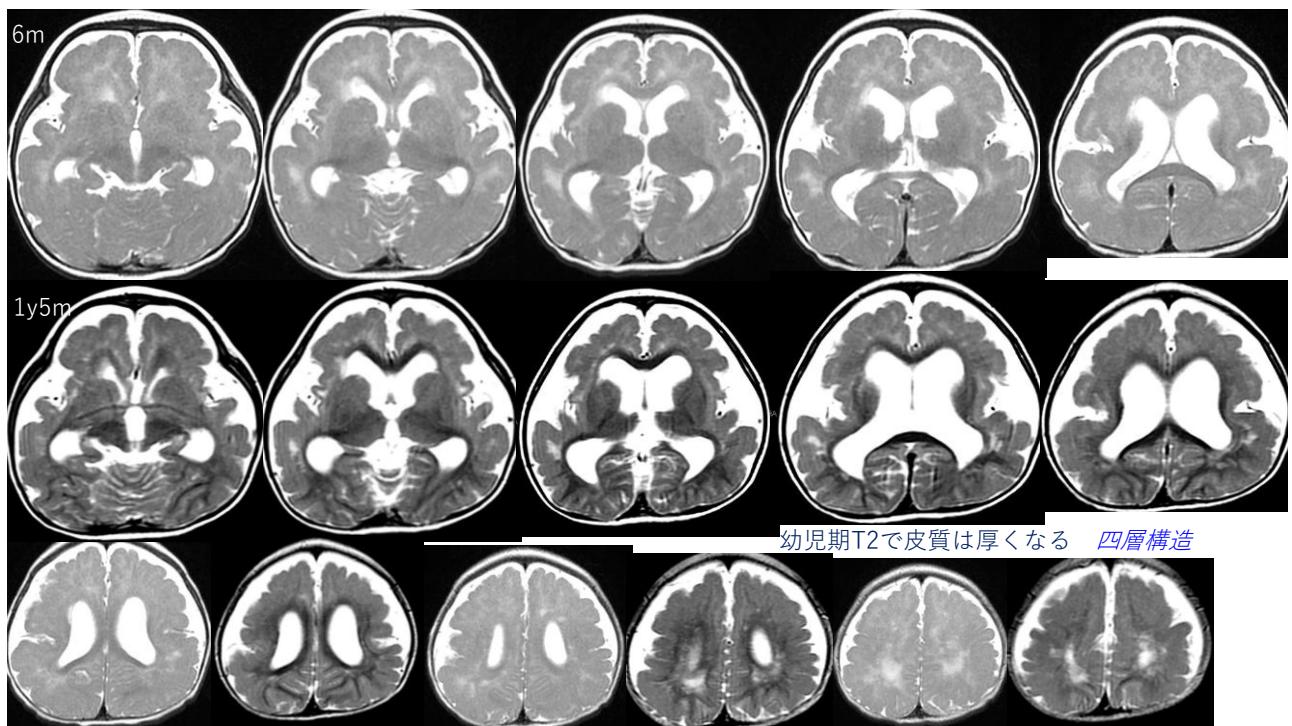
➤ PMGは病理学上定義されたが、現在は神経画像(MRI)上の概念である

- 病因は多彩であり(遺伝子・感染・外傷・循環障害…)、なお未確定が多い
- 脳軟膜・脳表の異常が多い *この部が脳回形成の主役かもしれない
- 過早期脳回形成の可能性もあり
- Cortical layerによる分類はartificialである *2層と4層の共存が多い
- Cortical fusionはないことが多い
- 正常皮質から異常皮質に突然変わることも多い
- Grey-white matter junctionが明瞭でないことが多い
- 他の奇形を合併することも多い

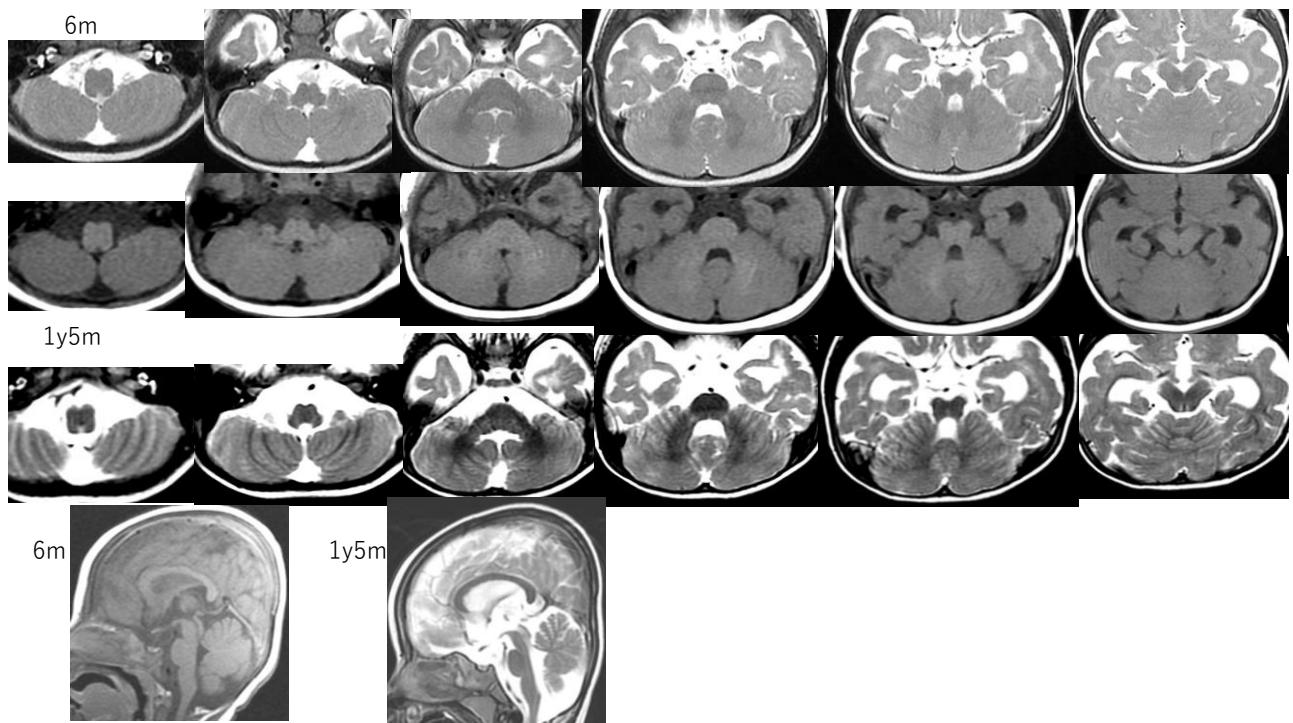
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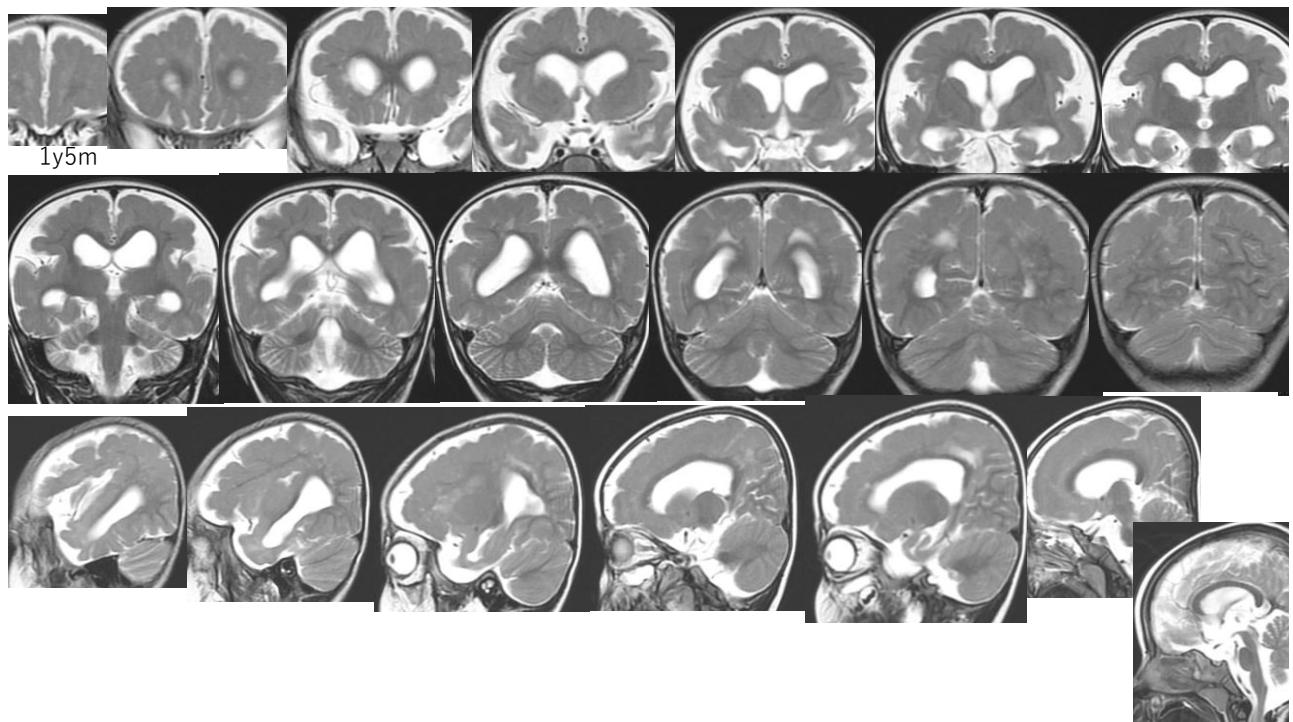
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32



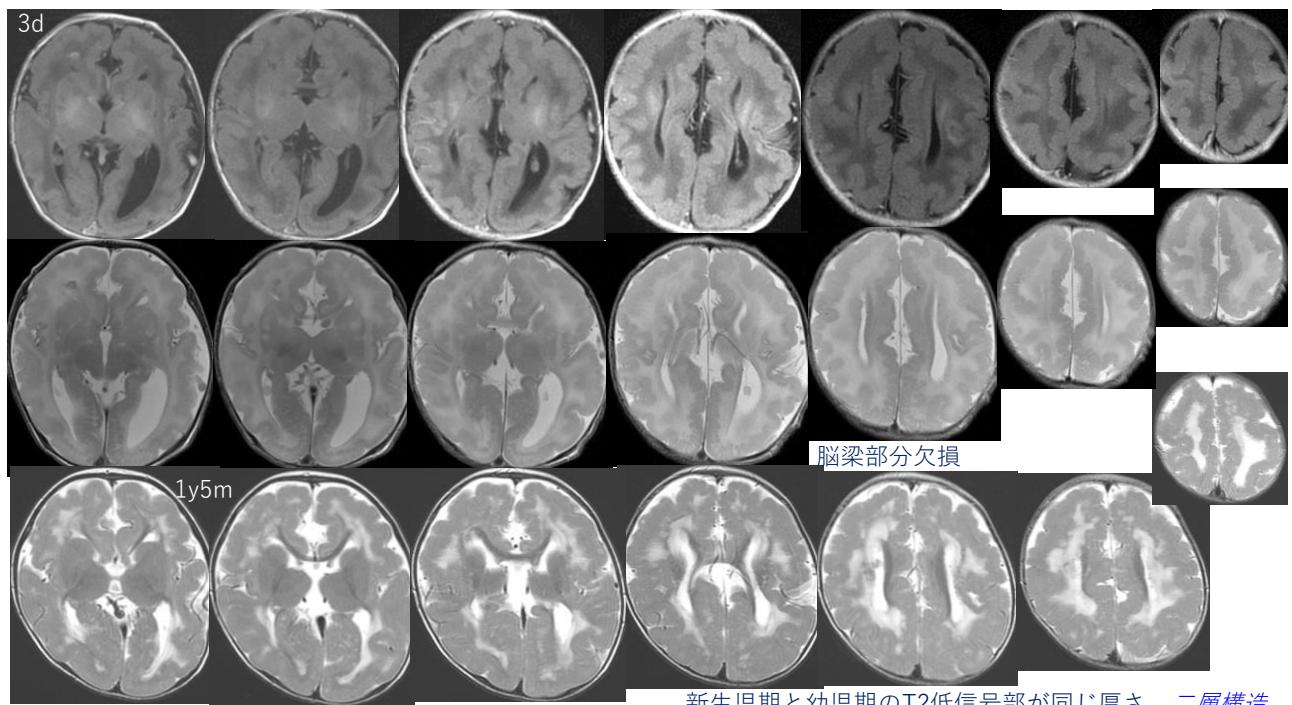
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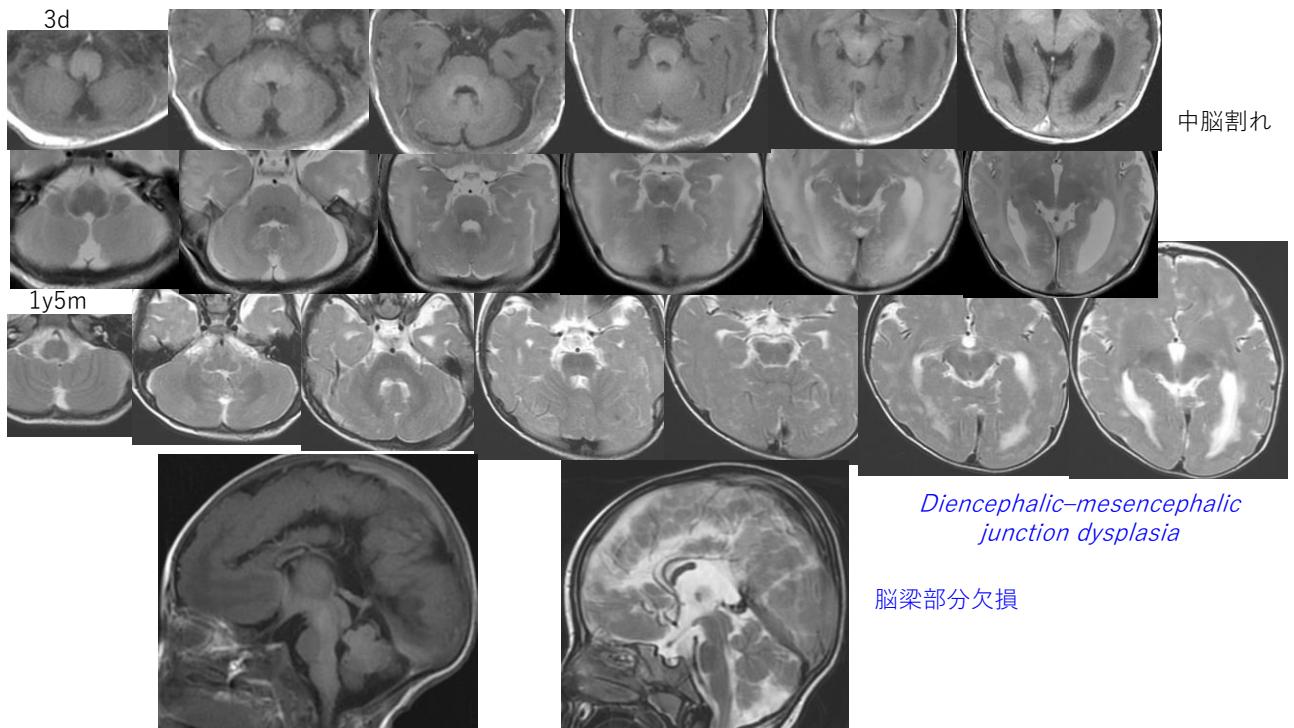
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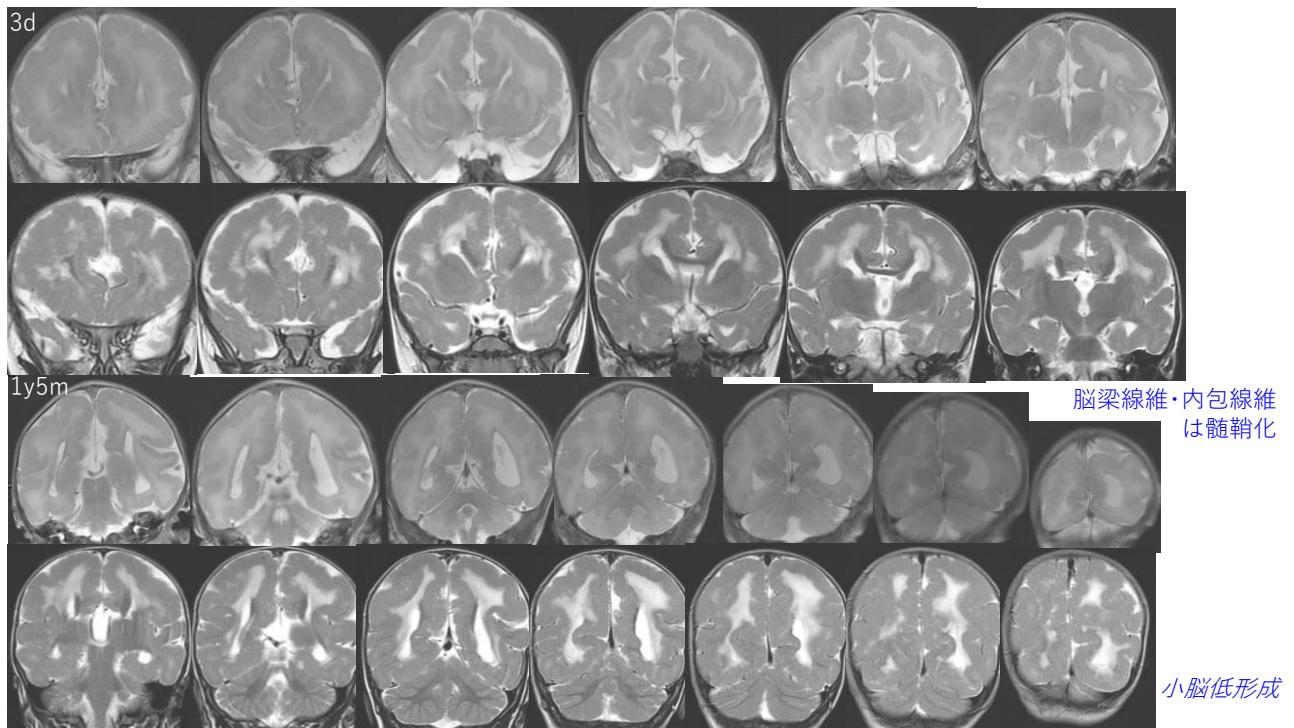
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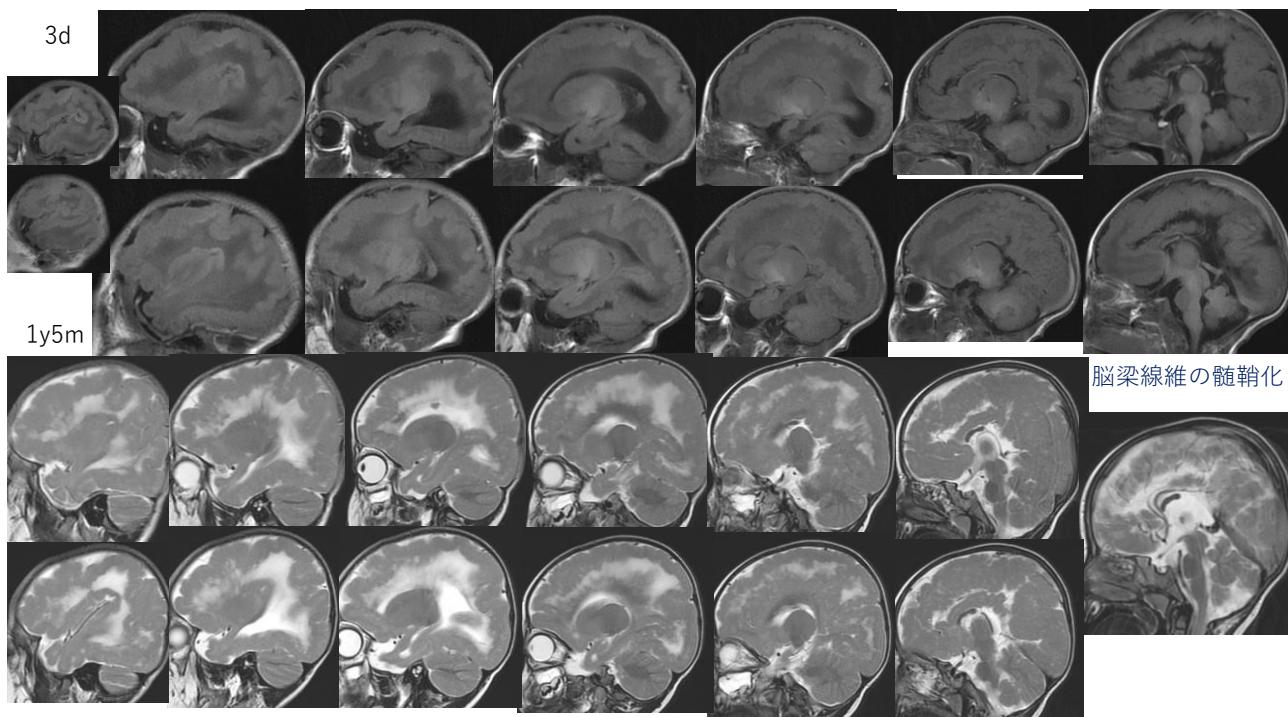
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37



38



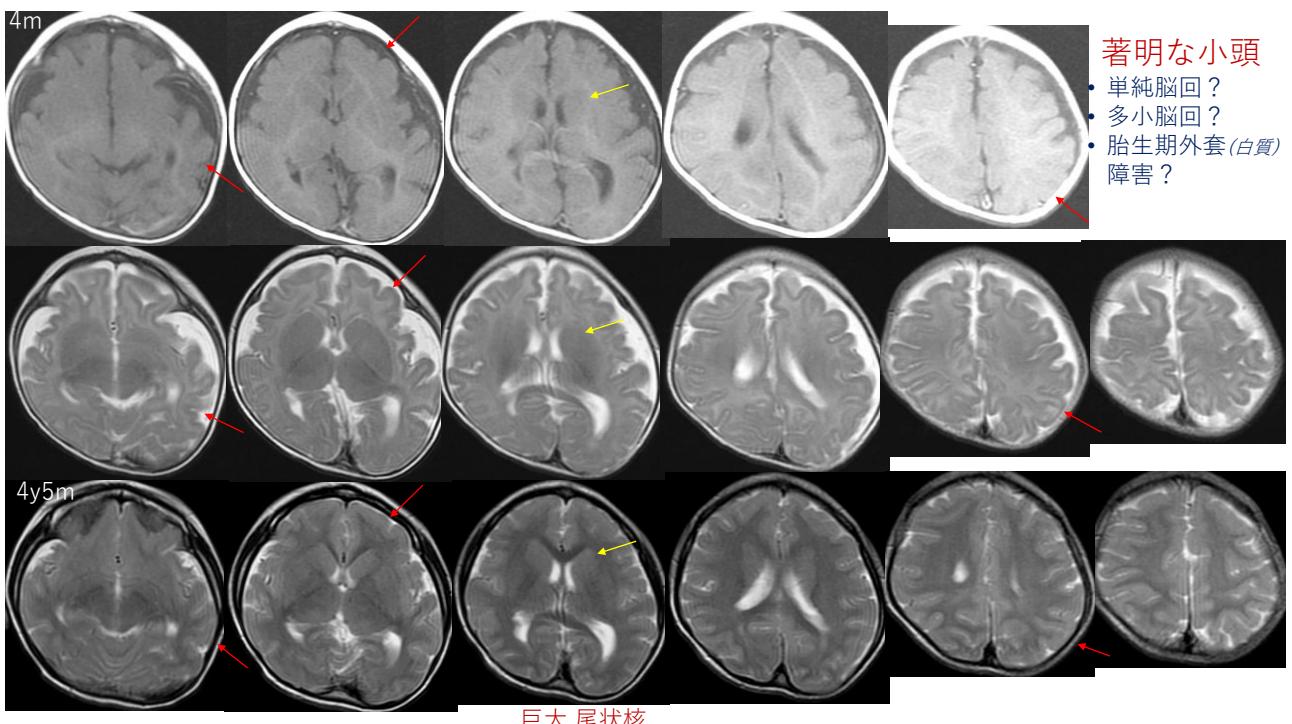
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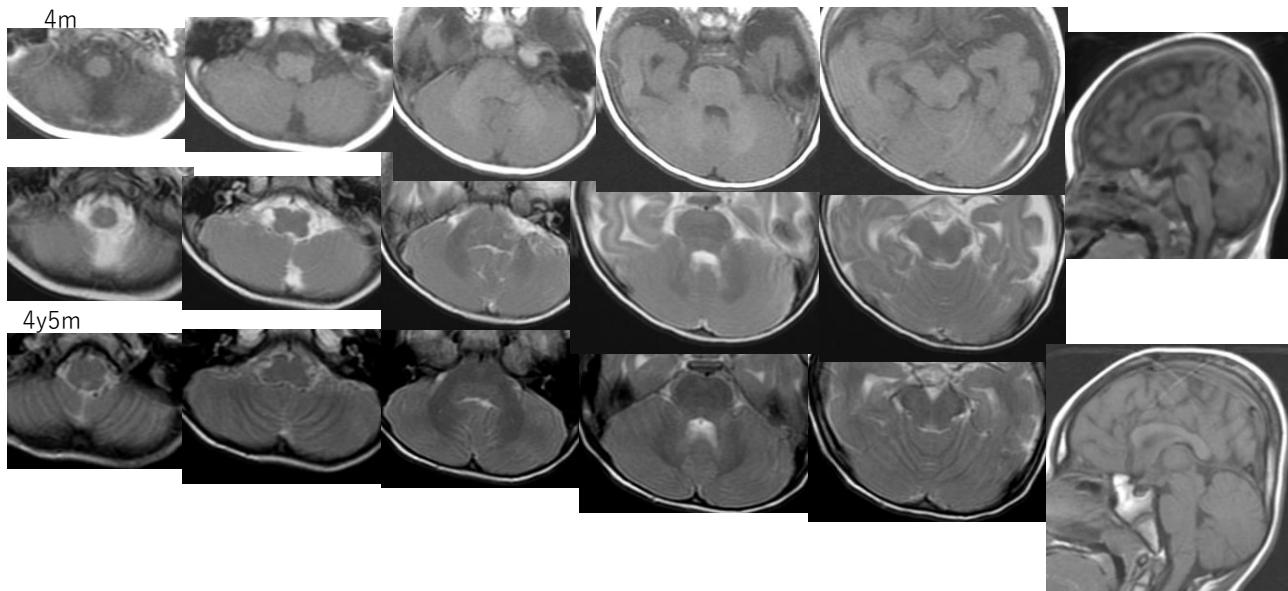
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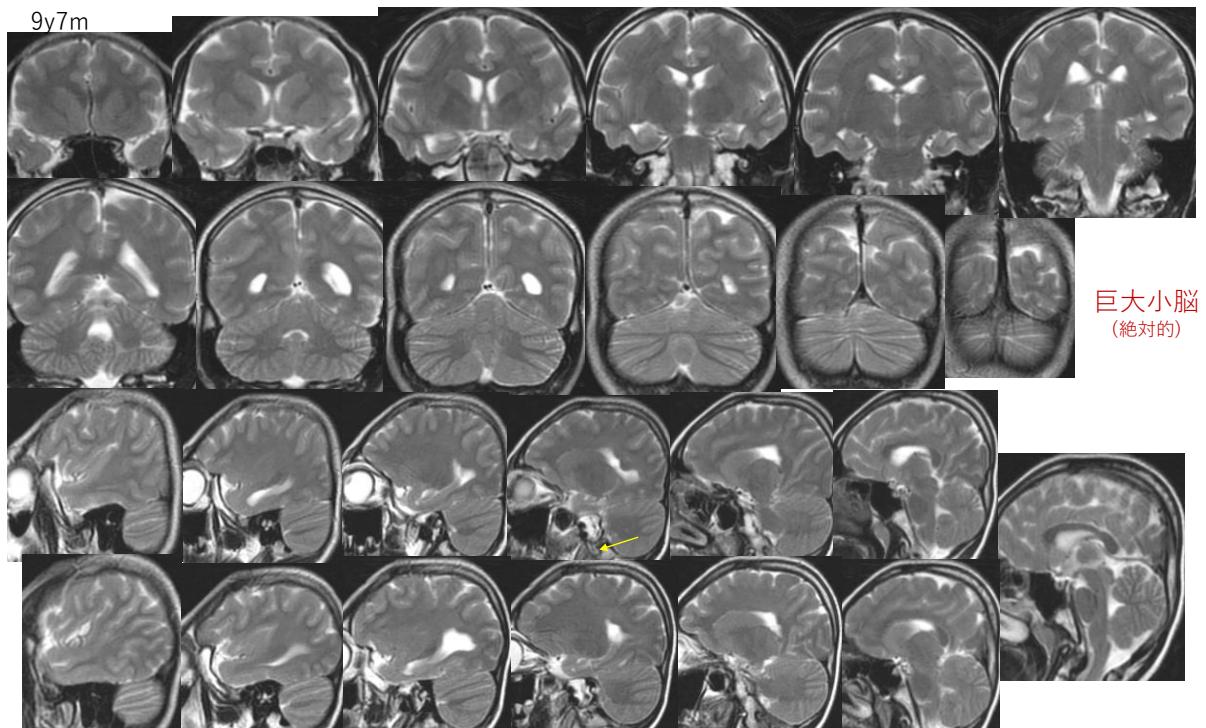
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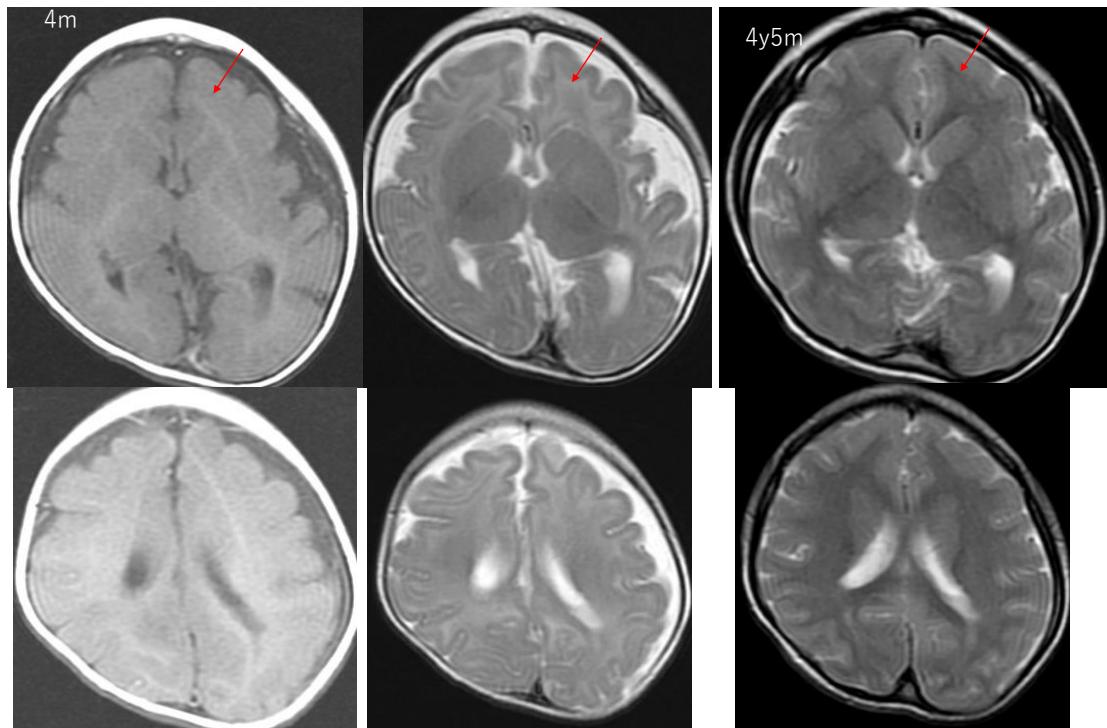
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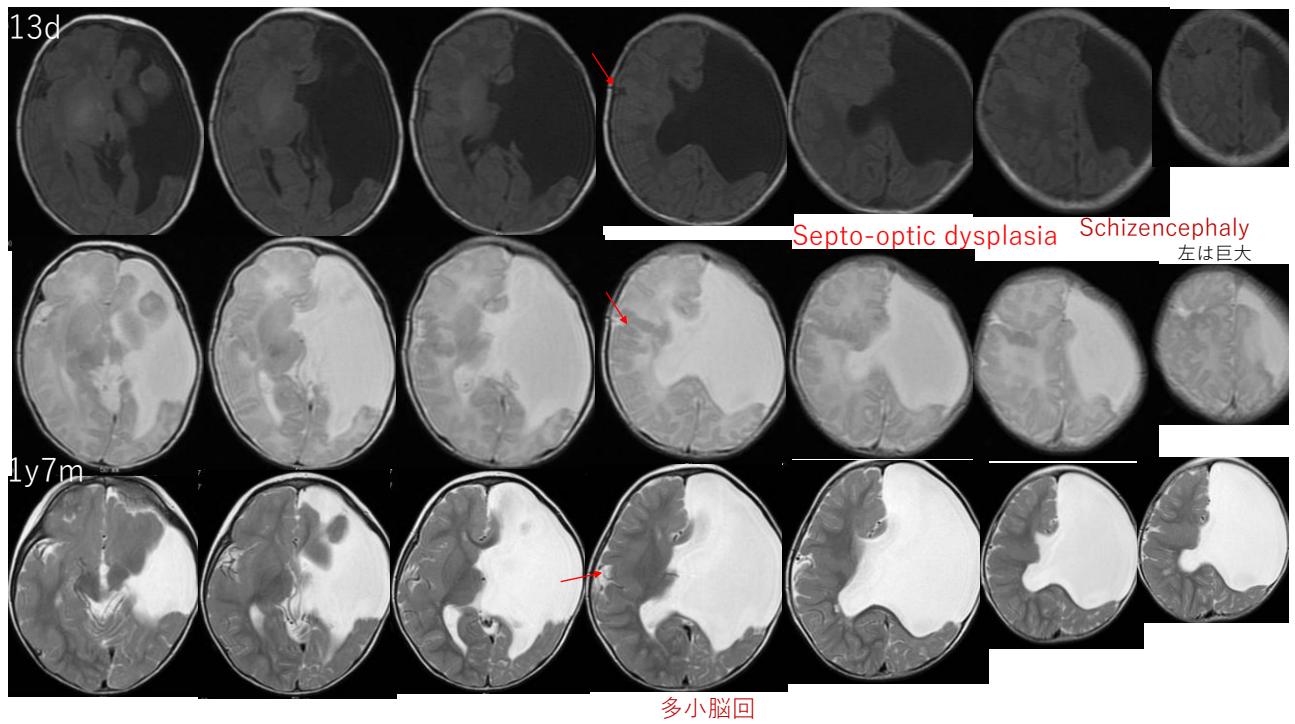
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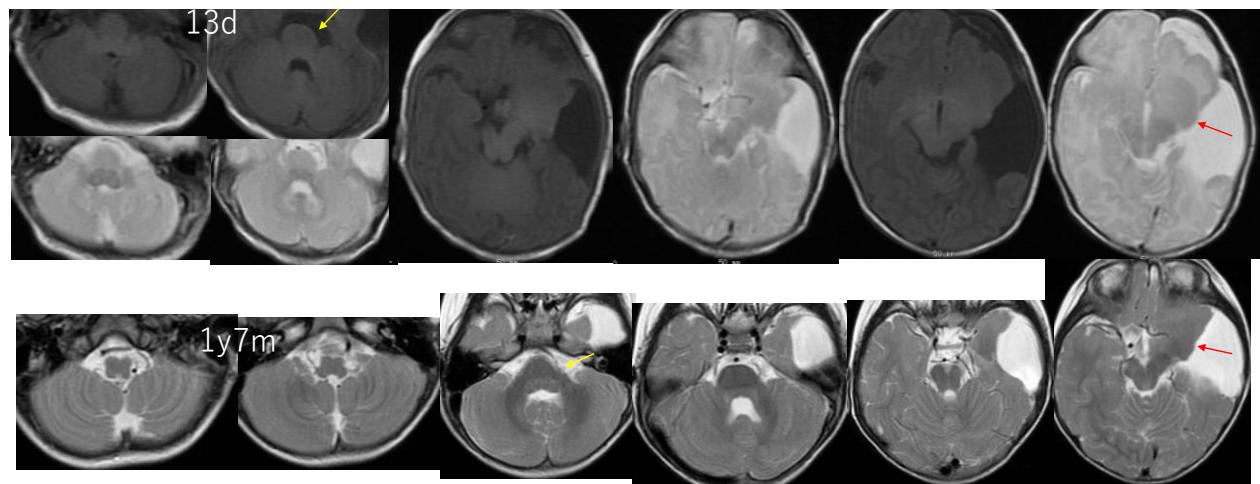
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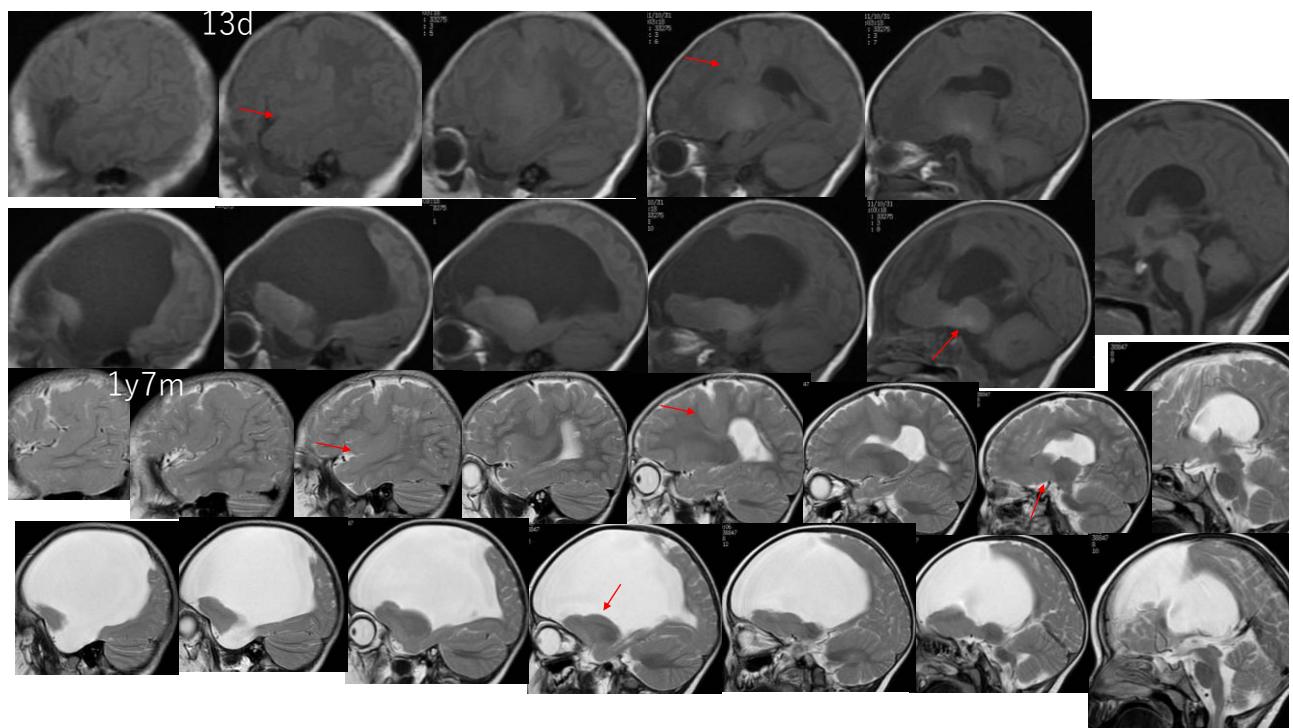
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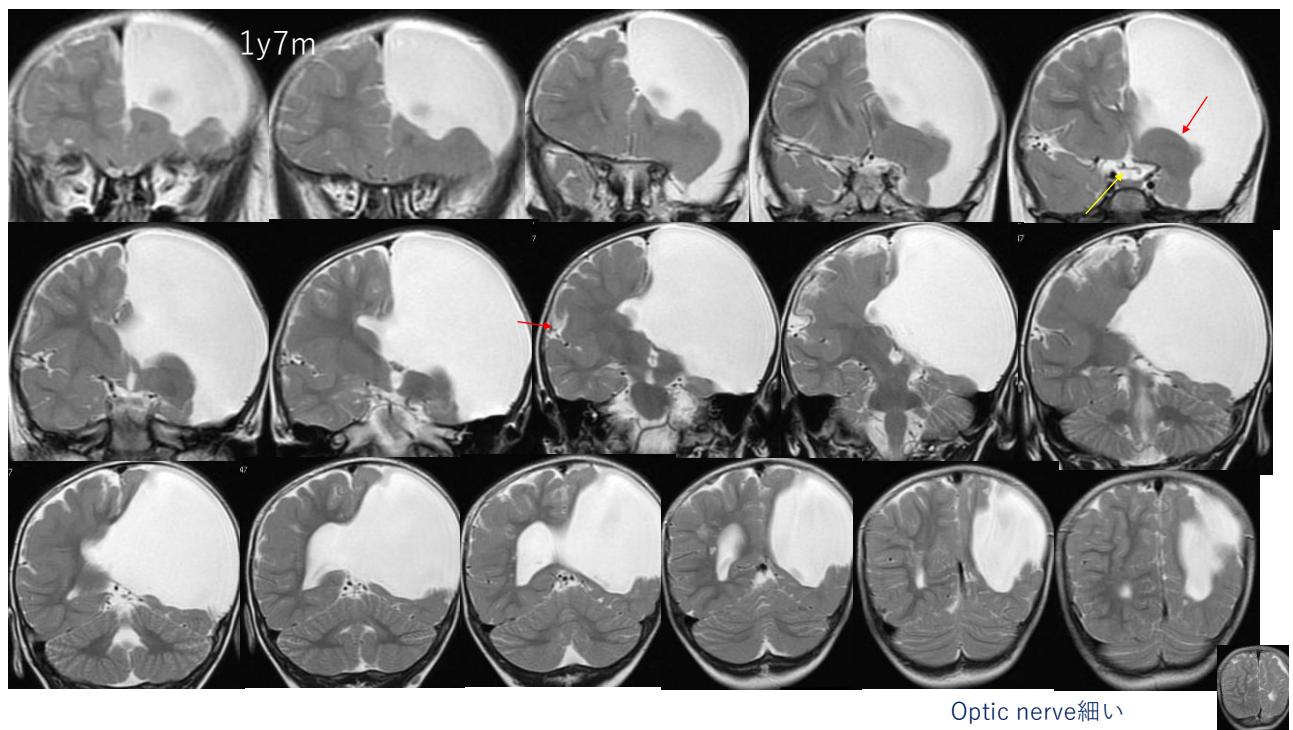
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48



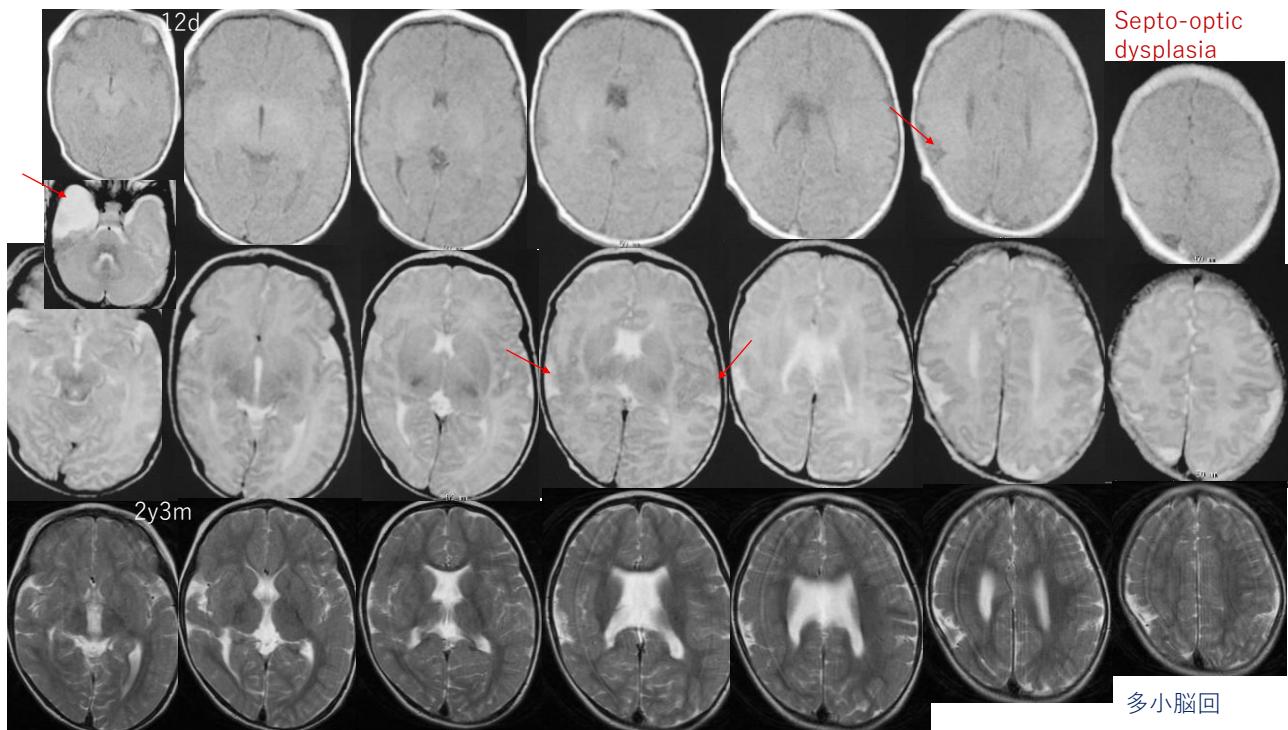
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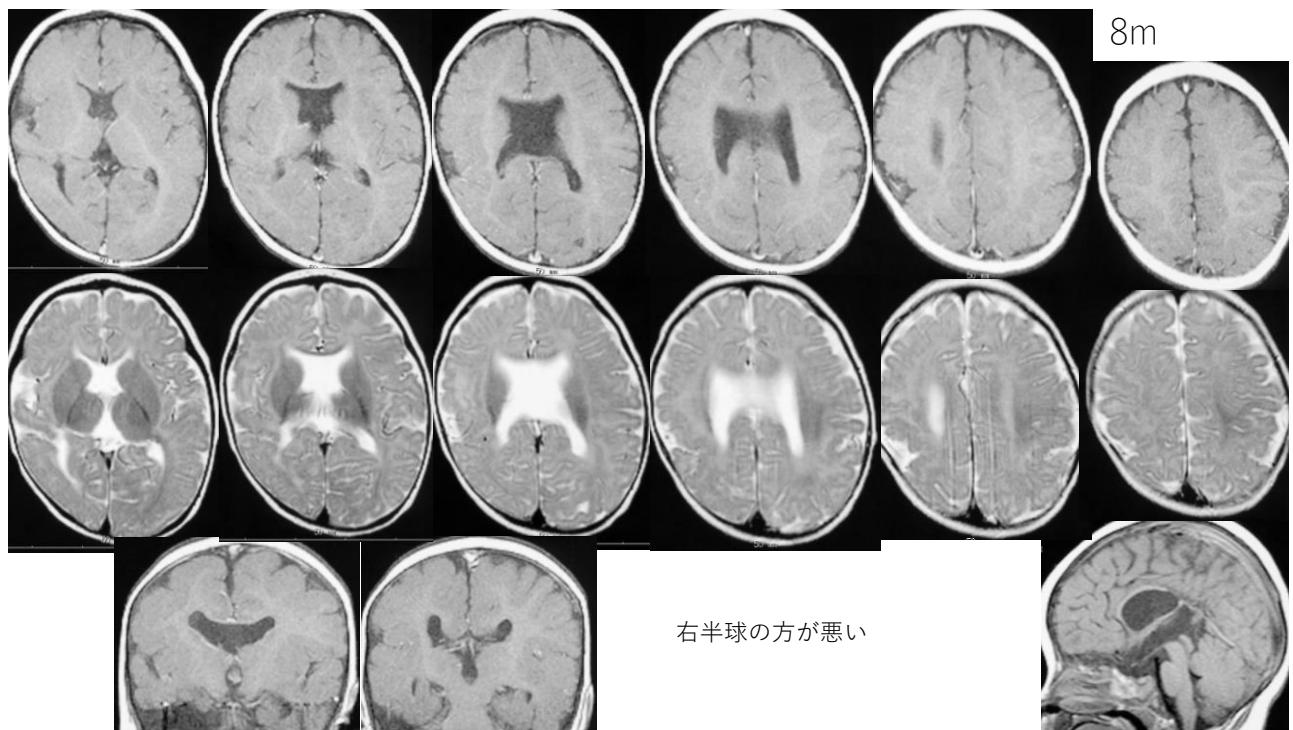
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51

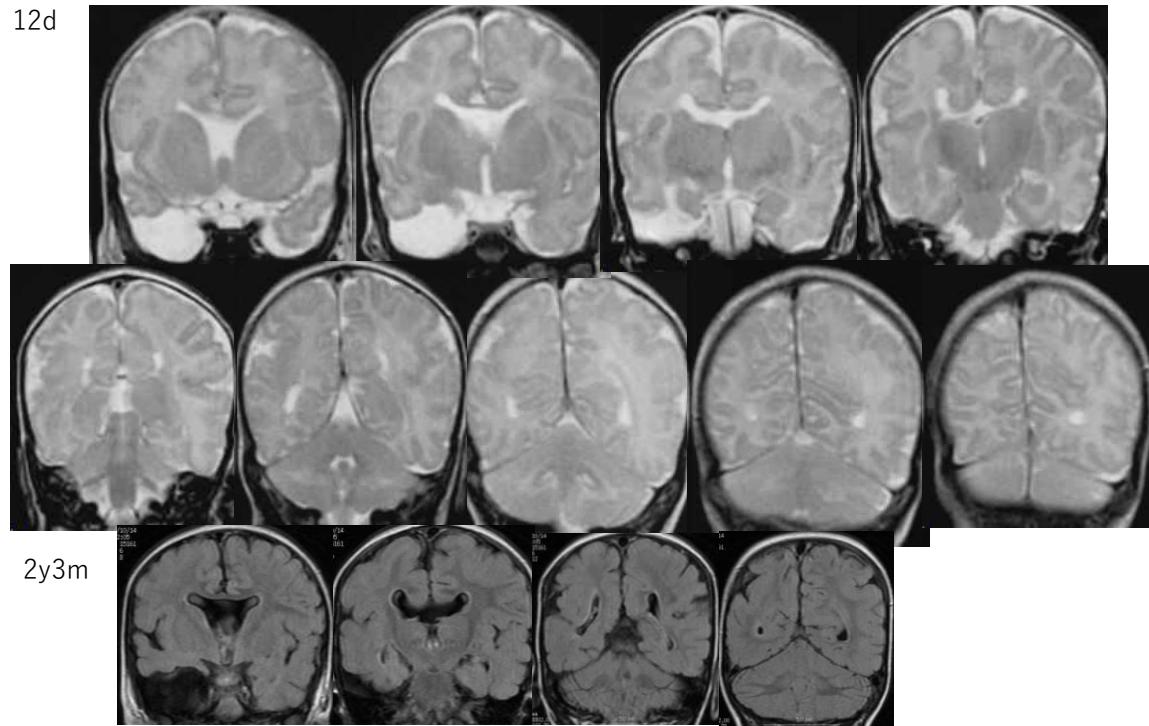


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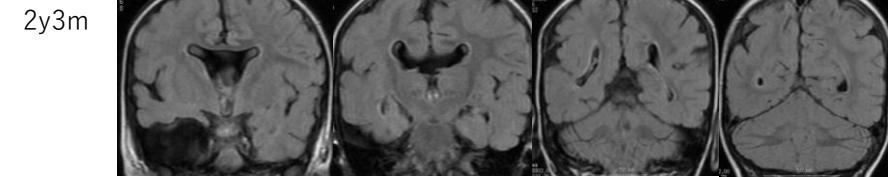


53

12d

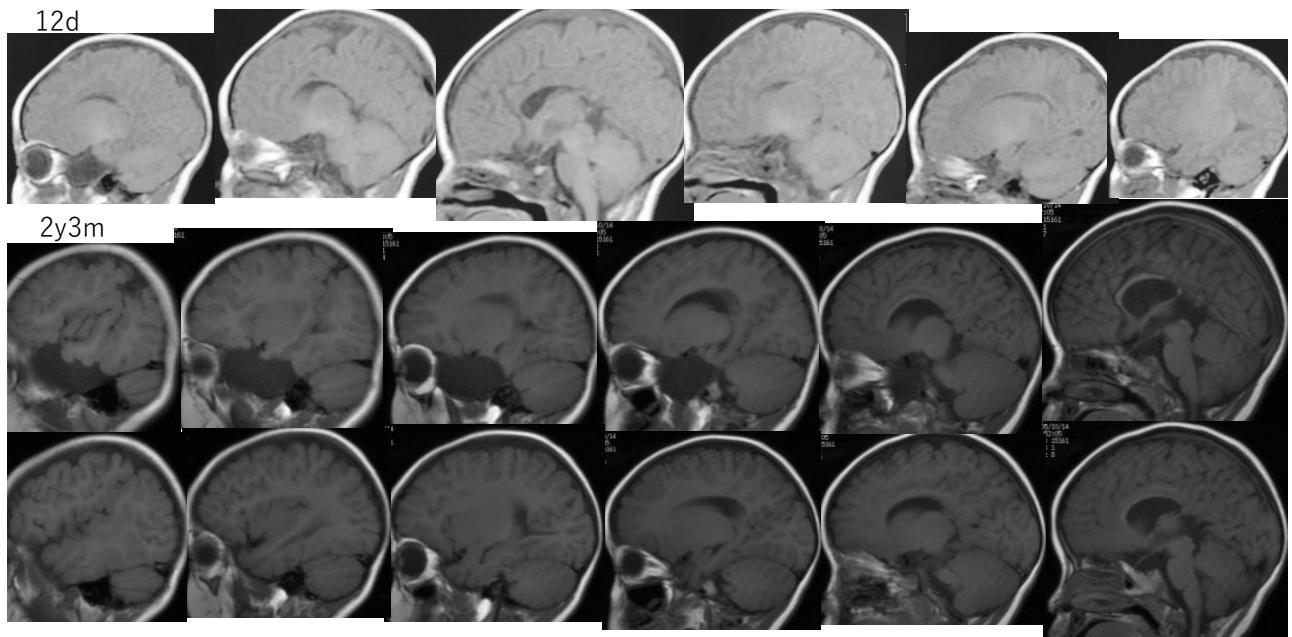


2y3m



54

右半球の方が悪い



55



56

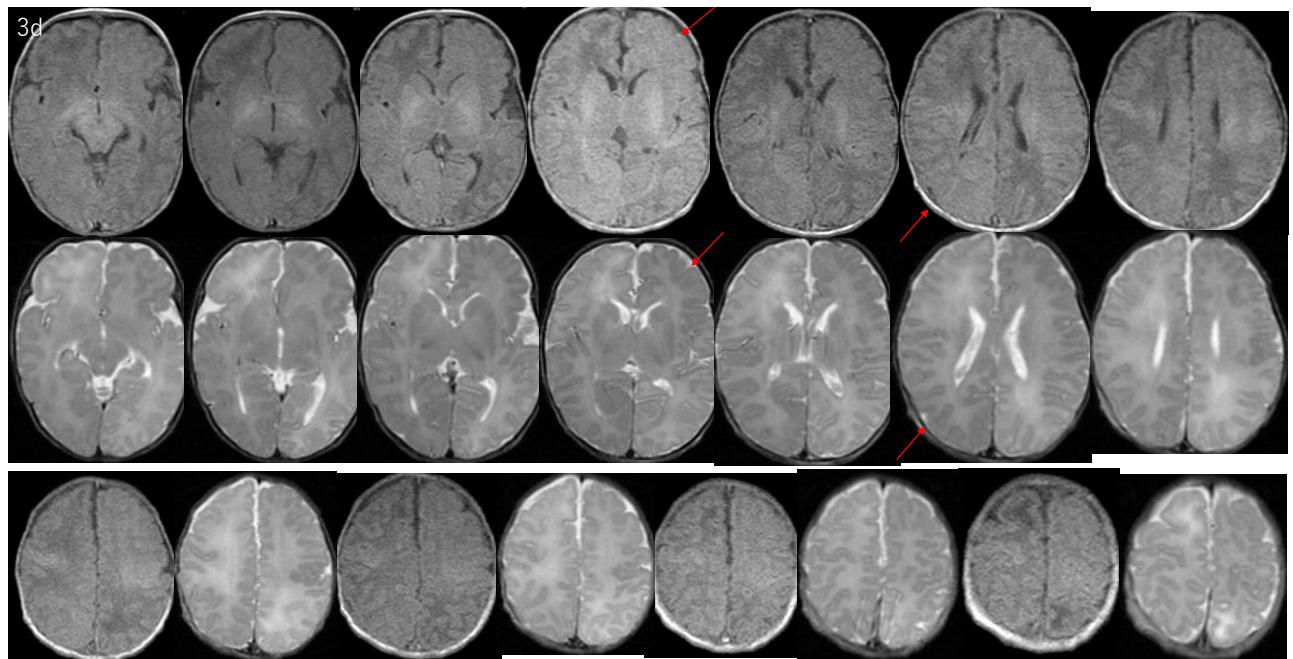


Paroxysmal ocular downward deviation
(Yokochi)

斜め上 ⇄ 斜め下 眼瞼と連動

【仮説】 中央視不能
↓
周辺視ネットワーク稼動
↓
Feedback不全でovershoot

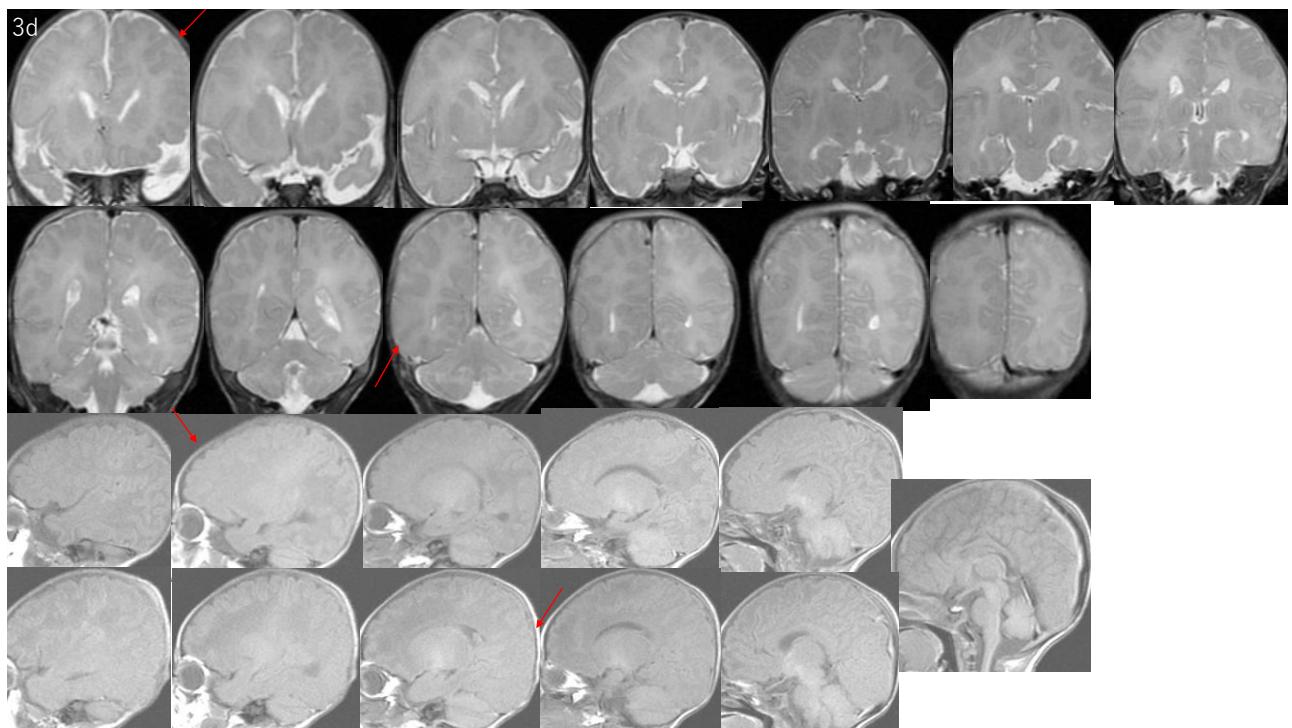
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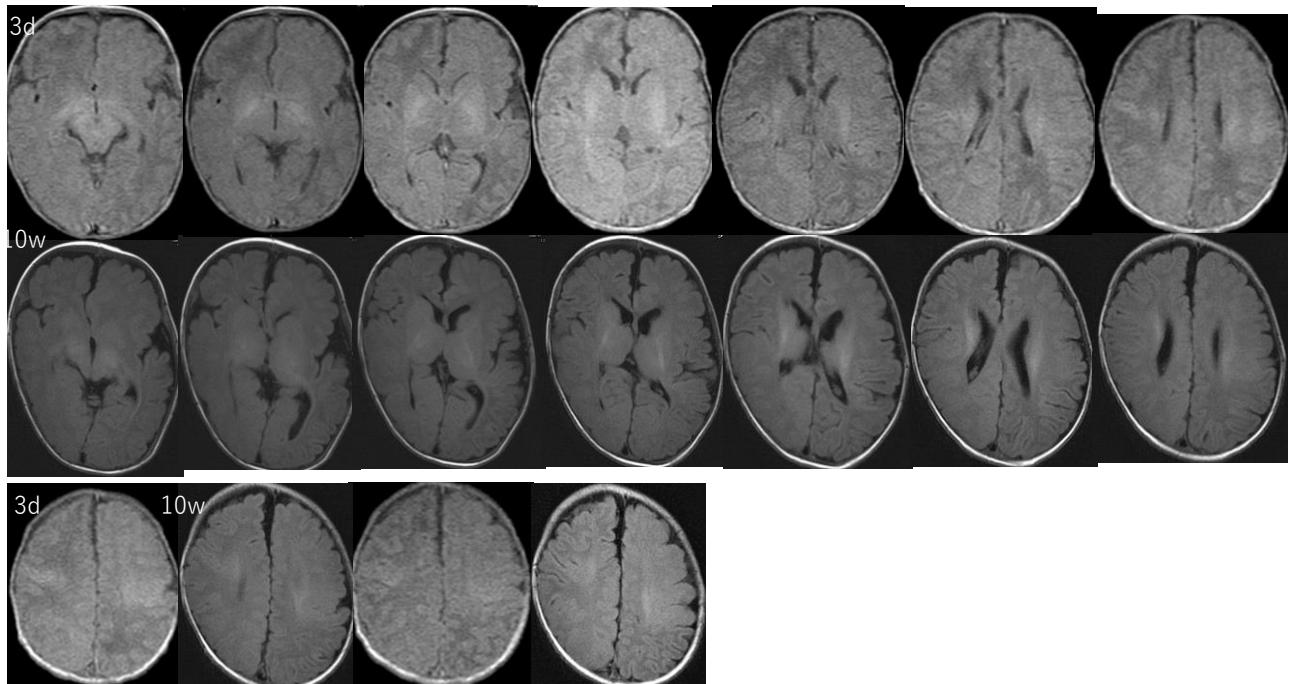
3dより痙攣頻発

局所的外套異形成

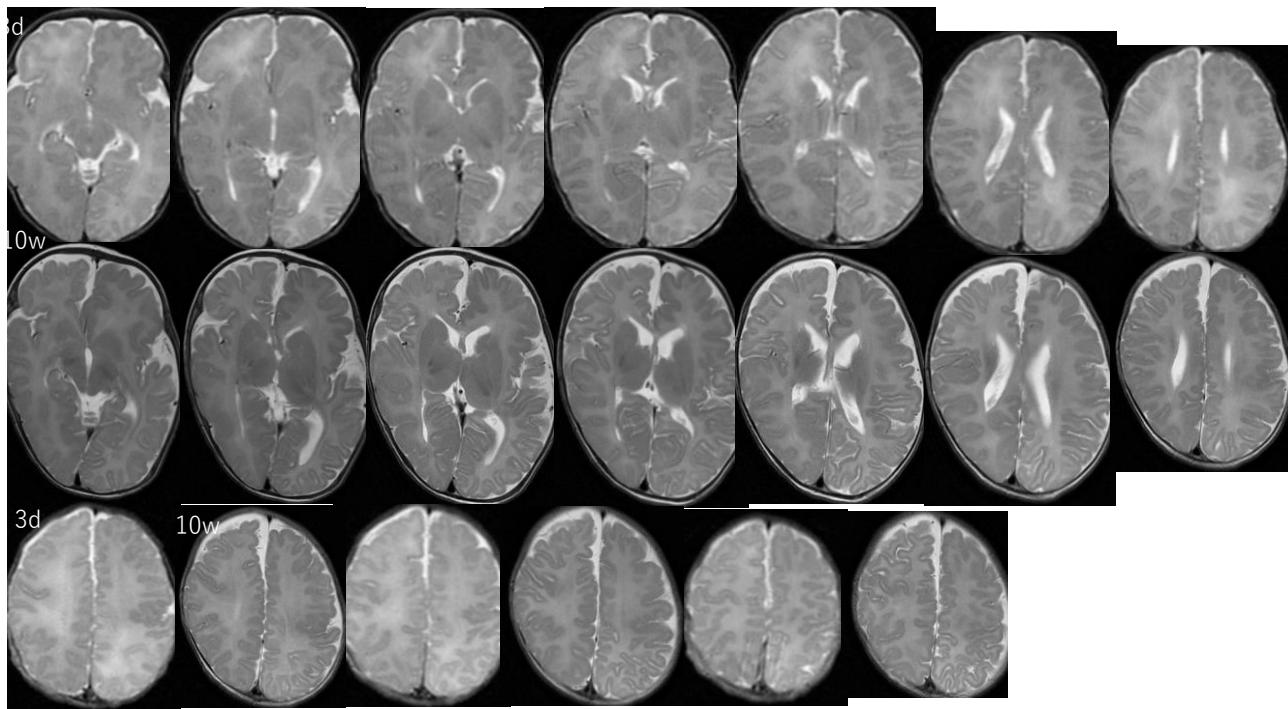
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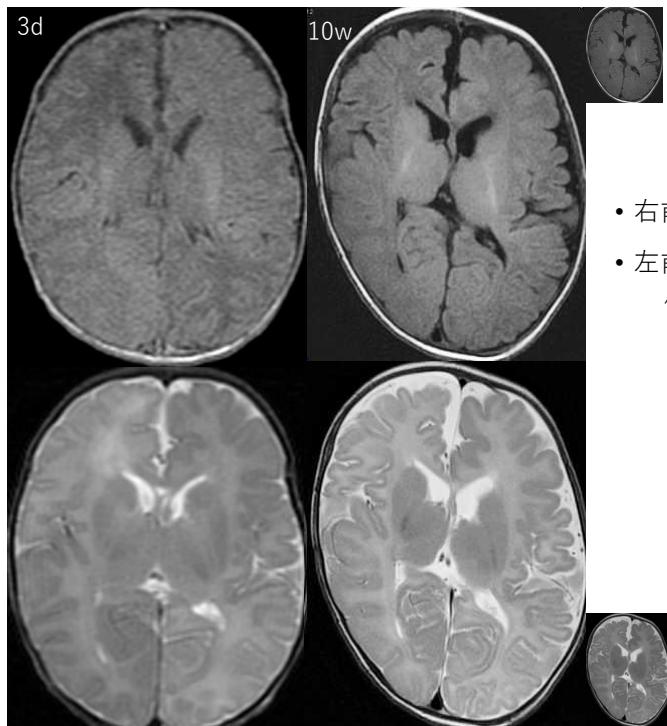
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前頭部白質

	3d	→10w
右	T1 黒 T2 白	灰 灰
左	T1 薄灰 T2 濃灰	薄黒 灰

- 右前頭部白質の信号変化は、髓鞘化に一致する
- 左前頭部白質の信号変化は、その逆
細胞過多部で、*apoptosis*による細胞減

外套分化障害

- 滑脳症
- 多小脳回
- Focal cortical dysplasia
- 髓鞘形成不全

脳回と白質の二分はだめ

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