

## WB-MRIの背景

# WB-MRI

## Pediatric Cancer Predisposition Imaging: Focus on Whole-Body MRI

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Cancer predisposition syndromeの患児

1. 若年からの長期にわたる定期的な画像スクリーニングの必要性
2. がん遺伝子・がん抑制遺伝子異常による放射線感受性の問題

→放射線被曝は避けたい。

→画像スクリーニングのmodalityはUSかMRI

### WB-MRIのメリット

1. 放射線被曝がない
2. 広い撮像範囲

## 小児遺伝性腫瘍のWB-MRIによるスクリーニングの適応範囲

Syndrome	WB-MRI	Brain MR	Spine MR	NCAP WB	CAP WB + Neck MR
LFS	Q 12 mo from Dx	Q 12 mo from Dx (alternating Q 6 mo with WB-MRI unless under GA)			
NFI	Baseline scan between 16 and 20 years				
NF2 and schwannomatosis	Based on Sx and location	Add IAM Q 6-24 mo	Q 24-36 mo		
RB	Q 12 mo from 8 y	Q 6 mo, if possible	Q 6 mo, if possible		
CHMRD (+/- LS)	Q 12 mo from 6-8 y	Q 6 mo, to 5 y	Q 6 mo, to 5 y		
DICER1 syndrome (no anaesthesia)	Consider	Q 6 mo, from Dx			
RTS	Consider	Urgent if Sx			
HPP syndrome				Q 24 mo from 6-8 y	Q 24 mo from 6-8 y

### \*WB-MRIの適応(recommendation)のない症候群もあることに注意!!

NOT INDICATED  
 Adenomatous polyposis syndromes (APC, MUTYH); juvenile polyposis coli (BMPRIA, SMAD4, PTEN); Peutz-Jeghers syndrome (STK11/LKB1); RA-Sopathies; NS; NSLAH; NSML; CFCS; CS; LS; CBL3; Sotos; Weaver; Rubinstein-Taybi; Schinzel-Gleason; and NKX2-1 syndromes; metabolic disorders linked to childhood cancers; rare DNA repair disorders; ataxia telangiectasia; Bloom syndrome; Fanconi anemia; dyskeratosis congenita; Nijmegen breakage syndrome; xeroderma pigmentosum; PHTS; HLRC syndrome; Gorlin syndrome; leukemia/lymphoma syndromes unless part of other syndrome, e.g., LFS; CHMRD; overgrowth disorders; and kidney tumors, e.g., BWS; DGS; FS; HB; IHH; SGBS; PROS; PS; WT; NB; MEN syndromes: MEN1, MEN2A and B, MEN4; HPT-JT; vHL disease

Abbreviations: BOS, Bohning-Ortiz syndrome; BWS, Beckwith-Wiedemann syndrome; CBL3, CBL syndrome; CFCS, cardiofaciocutaneous syndrome; CHMRD, constitutional mismatch repair deficiency syndrome; CS, Costello syndrome; DGS, Denys-Drash syndrome; Dx, diagnosis; FS, Fraser syndrome; GA, general anaesthesia; HLRC, hereditary leiomyomatosis and renal cell cancer syndrome; HPT, hereditary parathyroidism and pleurothoracicostoma syndrome; HPT-JT, hyperparathyroid-jaw tumor; IAM, internal auditory meati; IHH, isolated hemihypertrophy; LS, Legius syndrome; LS, Lynch syndrome; MEN, multiple endocrine neoplasia; NB, hereditary neuroblastoma; NF1, neurofibromatosis type 1; NF2, neurofibromatosis type 2; NS, Noonan syndrome; NSLAH, NS like with loose anagen hair; NSML, NS with multiple lentiginos; PHTS, PTEN hamartoma tumor syndrome; PROS, PIK3CA-related overgrowth spectrum; PS, Perlman syndrome; RB, hereditary retinoblastoma; RTS, Rothmund-Thomson syndrome; SGBS, Simpson-Golabi-Belmei syndrome; Sx, symptoms; WT, Wilms tumor; vHL, Von Hippel-Lindau; O, every; mo, months; y, years.

## WB-MRIの技術的側面

WB-MRIのスタンダードのプロトコールは存在しない。

各施設の放射線科の状況、MR装置のスペック、許容撮影時間、疾患の種類、患児の体格や年齢、技師を含む放射線科の人員、放射線診断医・診療科の医師の好みなどにより変化する。

MR装置： 1.5T vs 3.0T

年齢や体格によって異なるので、一概にはいえないが、一般論としては3Tの方が、DWI(拡散強調画像)では高磁場による磁化率アーチファクトがやすい。しかし、近年は小児のWB-MRIにおいて3Tの画質に問題ないという報告もでている。

撮像体位： 臥位（上肢は体の横）

撮像時間： さまざま（18分～72分まで報告がある）

鎮静： 6歳未満で考慮が必要。状況によって「feed and-wrap」プロトコール。全身麻酔の問題。Child life consultation/specialistの活用。

## 小児遺伝性腫瘍のWB-MRIによるスクリーニングの適応範囲

Syndrome	WB-MRI	Brain MR	Spine MR	NCAP WB	CAP WB + Neck MR
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NFI	Baseline scan between 16 and 20 years	Add IAM Q 6-24 mo Q 6 mo if positive	Q 24-36 mo Q 6 mo if positive		
NF2 and schwannomatosis	Based on Sx and location	Add IAM Q 6-24 mo Q 6 mo if positive	Q 6 mo from Dx		
RB	Q 12 mo from 8 y	Consider (no anesthesia)	Q 6 mo from Dx		
CHMRD (+/- LS)	Q 12 mo from 6-8 y	Consider (no anesthesia)	Q 6 mo from Dx		
DKER 1 syndrome	Consider	Consider	Urgent if Sx		
RFS	Consider				Q 24 mo from 6-8 y
HPP syndrome	Consider				Q 24 mo from 6-8 y

\* CAP WB-MRI: Chest, Abdomen and Pelvis MRI

\* \* NCAP WB-MRI: Neck, Chest, Abdomen and Pelvis MRI

基本的にWB-MRIといえば、「頭」〜「足」まで

しかし、症候群によって、それらの範囲を変えたり、WB-MRIとは別に局所領域のMRIを撮像することも考慮される必要がある。

撮像範囲が広い = 空間分解能の低下・撮像時間の延長  
撮像範囲が狭い = 空間分解能の向上・撮像時間の短縮

## WB-MRIの撮像プロトコル一覧 (論文から)

Authors and reference	Imaging plane	Sequences	Approximate scan time
Davis et al. (6)	Coronal	STIR, HASTE, T1 Option: MRA-TWIST	N/A
Eutsler and Khaana (7)	Axial	STIR	40 minutes
Nevelstein and Litoolj (8)	Coronal	STIR-HASTE, DWI (b values: 50, 400-500, 800-1000 $\mu\text{m}^2$ )	32 minutes
Viliani et al. (17)	Axial	STIR, T1 T2 SE DWI-STIR (b values: 0, 100, 800-1000 $\mu\text{m}^2$ ), T2 SPAIR	18 minutes
Anupindi et al. (18)	Coronal	STIR, T1, HASTE	AVG 72 minutes
Jaspersion et al. (19)	Axial	STIR (head, neck, lower extremities), T2 FS (chest, abdomen $\pm$ pelvis), HASTE	< 1 hour

Abbreviations: AVG, average; DWI-STIR, DWI with background body signal suppression (DWIBS), applying prepulse of STIR for fat suppression; FS, fat suppression; HASTE, ultrafast half-Fourier-acquired single-shot turbo spin echo; MRA, magnetic resonance angiography; MRA-TWIST, dynamic time-resolved MRA; N/A, not available; SPAIR, spectral attenuated inversion recovery; STIR, short tau inversion recovery; T2 SE, turbo spin echo.

\* Approximate scan times vary with number of stations, determined by patient height, e.g., average of 5; authors 6-8, 16-18 (see references).

ただし、症候群によってWB-MRIだけでなく、局所MRI(頭部MRIなど)を加える方がよい。とくにCMRDS(constitutional mismatch repair deficiency syndrome), HB(hereditary retinoblastoma), LFS(Li Fraumeni syndrome)

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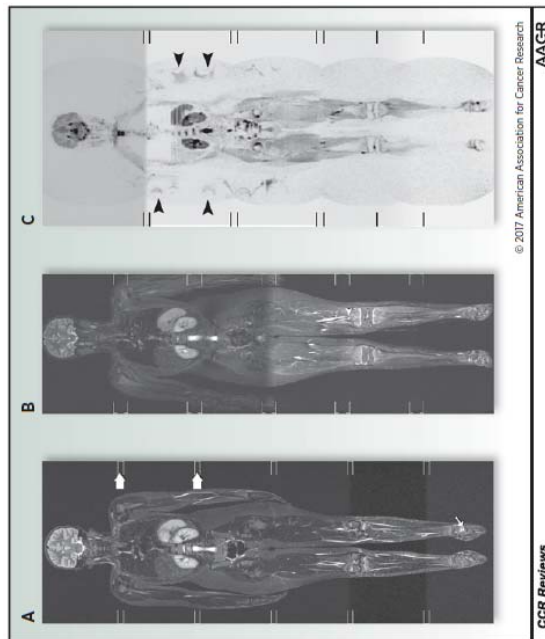
最も頻用されているシーケンス

1. 冠状断のFluid-sensitive 2D-sequence  
→主にSTIR(Short tau inversion recovery)

2. WB-DWI(拡散強調画像) “DWIBS”

\*造影剤は通常使用しない。

## Li-Fraumeni syndromeの女兒の毎年のWB-MRIによるスクリーニング



CCR Reviews

9歳

10歳

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AAGR

# WB-MRIの読影とレポートについて

## 1. どこまで小さい病変をdetectできるのか？

WB-MRI vs PET/CT: 12mm以下の骨浸潤はWB-MRIがよい。  
(Niveaustein RA et al. Radiol Med 2016)

肺結節については、4-10mmのものはWB-MRIでも高い感度で検出可能。  
ただし、CTがあくまでreference standard。

(Smith EA et al. Pediatr Radiol 2016)

## 2. 誰が読影し、レポートを発行するのか？

WB-MRIの読影に精通し、Cancer predisposition syndromeの知識を持つ放射線診断専門医

しかし、施設等の状況により、それは一定しない。

- ・一般放射線科医 vs 各領域を専門とする放射線科医(神経放射線科医・腹部放射線科医)
  - ・全身読影を一人の放射線科医が行い、その後各領域の専門放射線科医がdouble checkする
- 議論があるが、いずれにしろある一定のフォーマットに沿ってレポートを作成すべき。

## WB-MRIスクリーニングにおける放射線診断レポートのテンプレート例

<p><b>Table 4.</b> Report template: WBMRI without contrast</p> <p><b>CLINICAL HISTORY:</b> Surveillance for [LFS]</p> <p><b>COMPARISON:</b> [None/prior study from]</p> <p><b>TECHNIQUE:</b> Coronal STIR, coronal T1, axial T2 FS, optional axial DWI</p> <p><b>FINDINGS:</b></p> <p><b>SUPPORT DEVICES:</b> [None]</p> <p><b>HEAD/NECK:</b></p> <p>[The visualized anatomic structures of the head and neck are grossly normal and no mass is identified. There is no lymphadenopathy in the neck or supraclavicular region. The thyroid gland has uniform signal and no evidence for nodule, cyst, or mass.]</p> <p><b>CHEST:</b></p> <p>[There is no axillary, hilar, or mediastinal lymphadenopathy. The lungs demonstrate no pleural effusions, focal nodules, or consolidation. Breast tissue is normal. No mass is seen.]</p> <p><b>ABDOMEN:</b></p> <p>[The liver, spleen, pancreas, adrenals, and kidneys appear normal in signal and morphology. There is no evidence for mass in the solid organs, bowel, or mesentery. There is no abdominal or retroperitoneal lymphadenopathy.]</p> <p><b>PELVIS:</b></p> <p>[Male: The bladder and prostate are unremarkable. There is no free fluid.] [Female: The uterus is normal in signal and morphology. The ovaries are not visualized/are visualized and appear normal. The bladder is unremarkable. Free fluid is physiologic.]</p> <p><b>BONE MARROW/BONES/SOFT TISSUES:</b> The bones are normal in morphology. There is no abnormal bone marrow signal or focal bone abnormality. No soft tissue masses are identified.</p> <p><b>IMPRESSION:</b></p> <p>[NOT EVIDENCE FOR SOLID TUMORS [INCIDENTAL FINDINGS: NONE/INCLUDE] [ADDITIONAL IMAGING IS/IS NOT RECOMMENDED.]</p>
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## WB-MRIスクリーニングにおける遺伝性腫瘍ごとの要チェック部位

Anatomic location	Pediatric CPS disorders and disease groups										
	Neurofibro- masas			Gastrointestinal cancers		Neuroendocrine tumors		Erythema and lympho- matoid and immunodeficiency disorders		Miscellaneous disorders	
	LFS	NFI	Sch	CHMRD (LFS)	HPP (PHEO and PGL)	RTS	DICER1	RB			
Brain	X	X	X*	X*				X			X
Spine			X								
Orbils		X									X
Thyroid	X*										X
Lungs	X*										X
Heart											
Parathyroid											
Liver											
Pancreas											
Adrenals	X	X			X						X
Kidneys	X*			X*							
Urinary bladder	X*			X*							X
Uterus	X*			X*							X
Ovaries	X*			X*							X
Prostate											
Testis		X		X*							X
Bowel	X	X									
Bladder	X	X									X
Bone	X	X									X
Soft tissue/muscle	X	X									X
Hem/BM*	X	X									X
Skin	X	X									X
Other	X	X		X*							X

NOTE: Dark grey columns: WBMRI optional X (bold), "core" tumors most closely linked to syndrome/disease; Hem/BM\* = hematologic; bone marrow/lymph nodes; LFS, Li-Fraumeni syndrome; X\*, Brazilian founder mutation, other = neuroblastoma, CHMRD, constitutional mismatch repair deficiency syndrome, LS, Lynch syndrome; X\*, genitourinary; X, common to Lynch syndrome and CHMRD, other = neuroblastoma, RTG, Retinoblastoma, DICER1, other = nasal chondrosarcoma/lymph hamartoma, HPP syndrome, hereditary pheochromocytoma (PHEO) and paraganglioma (PGL) syndrome, bowel, gastrointestinal stromal tumor (GIST), other = neck/upper mediastinum–parasympathetic; lower mediastinum/abdomen/pelvis—sympathetic nervous systems; RB, hereditary retinoblastoma; NFI, neurofibromatosis type 1; X\*, genitourinary, bowel—GIST; other = nerve sheath tumors; NF2, neurofibromatosis type 2; Sch; schwannomatosis; X\* = internal auditory meati, other = where symptomatic; PHEO, multiple endocrine neoplasia.

## 結論

WB-MRIは、Cancer predisposition syndromeの多くの患者のルーチンスクリーニング検査として有用であり、それぞれの症候群にあわせて最適なプロトコールで行う必要がある。

各施設でWB-MRIの使用が可能かどうかなどは、この論文の主旨を超えているので述べられないが、いずれにしても放射線科医にconsultationして最適なスクリーニング法を考えることは重要と考えられる。