Imaging in Neuroblastoma

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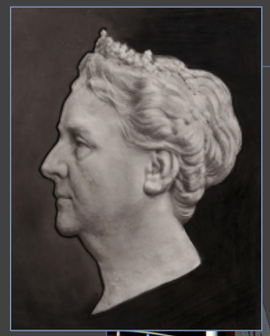
ESPR
European Society of Paediatric Radiology



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No disclosures

Content

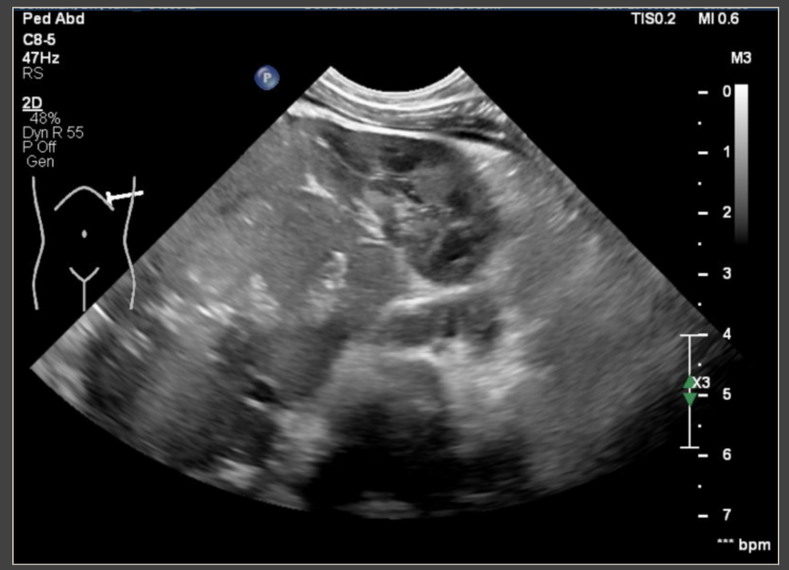
Introduction

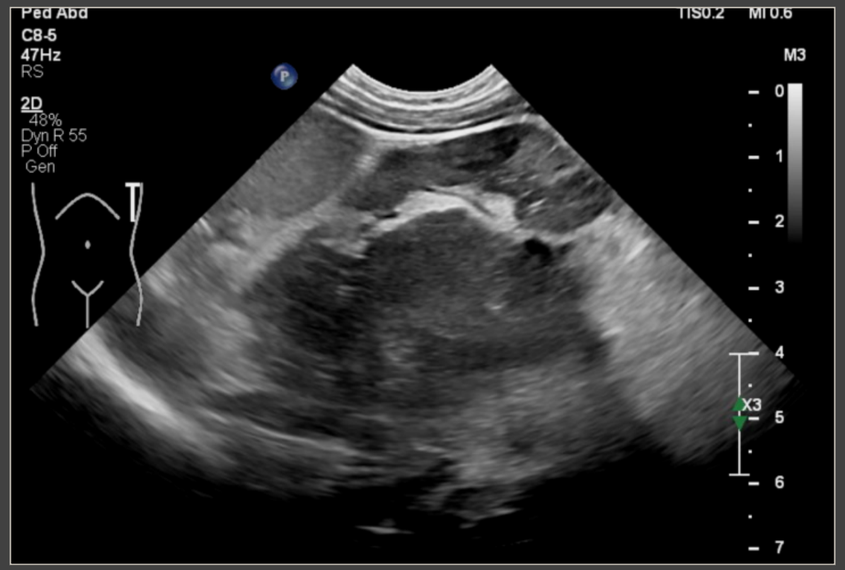
Imaging primary tumor - HR NBL 2 protocol

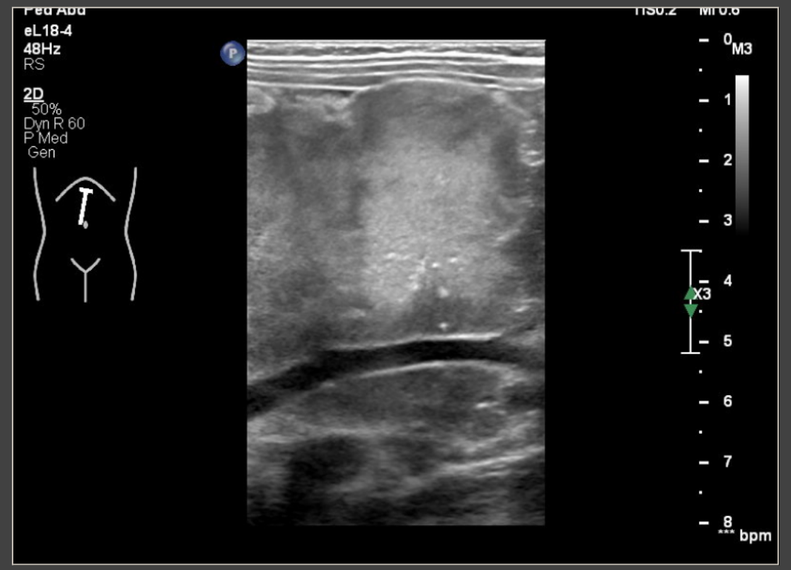
Distant metastases

Take home points

Case – 2-year-old boy. Not feeling well







Introduction

1. Location - Site of origin:

    Claw sign

    Movement

    Prominent vessel sign

2. Tumor characteristics (echogenicity, fat, calcifications..)

3. Patterns of spread

Key to differential diagnosis

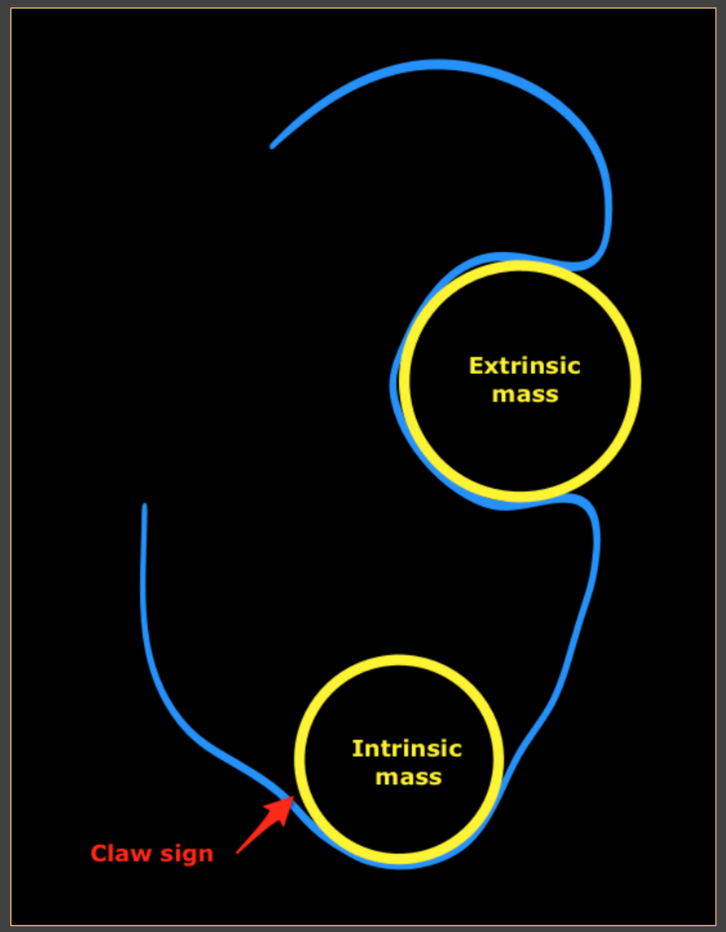
Imaging

- Location – Organ of origin
- Tumor characteristics
- Patterns of spread

Age

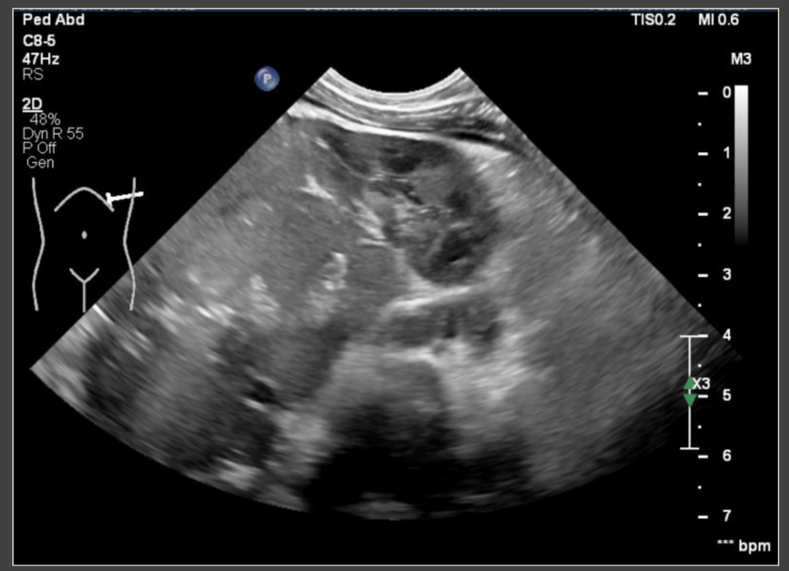
Tumor markers (urine metabolites)

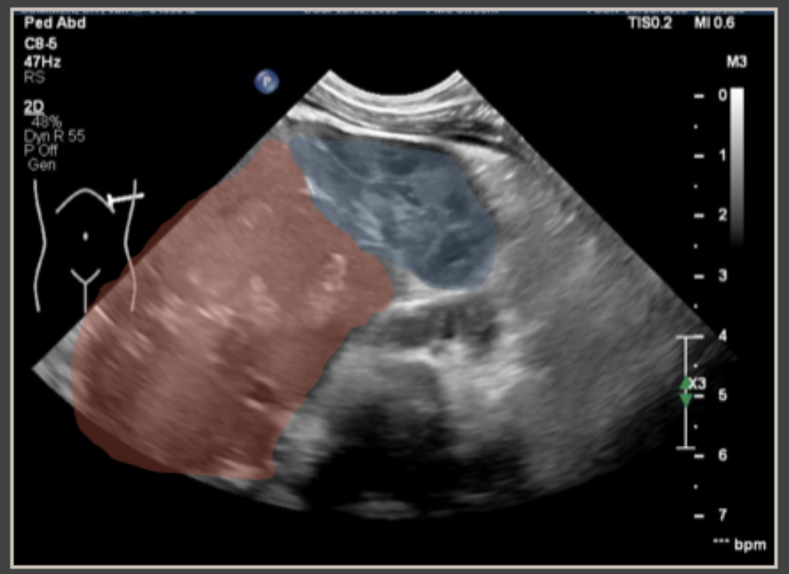
Claw sign

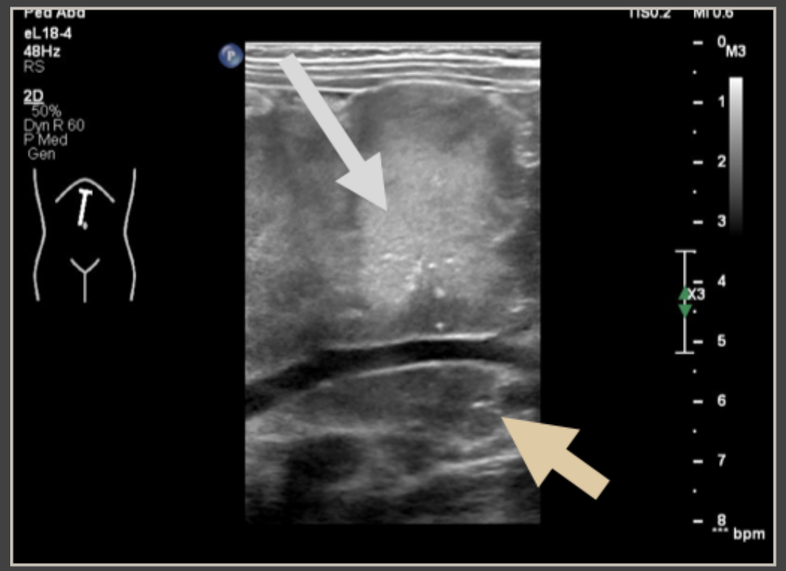


Case – Claw sign? Imaging characteristics?

Calcifications

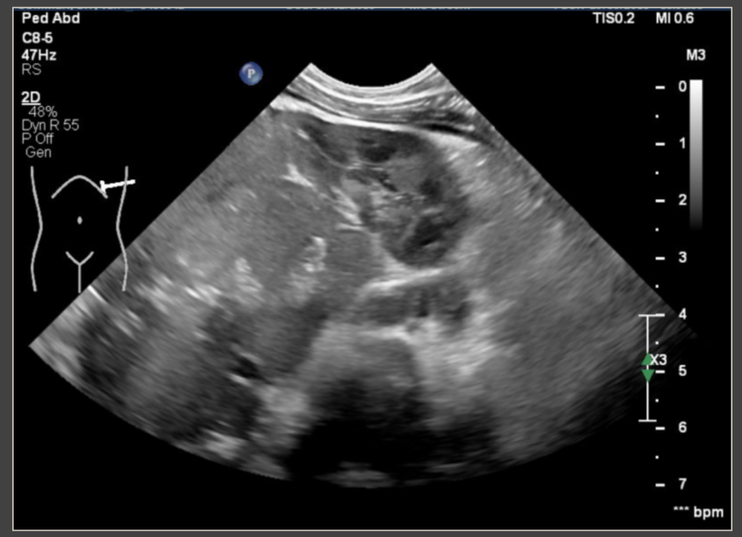


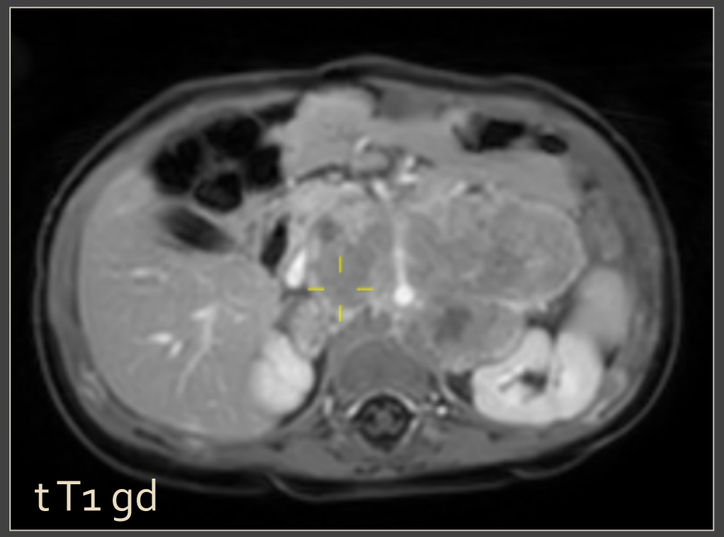


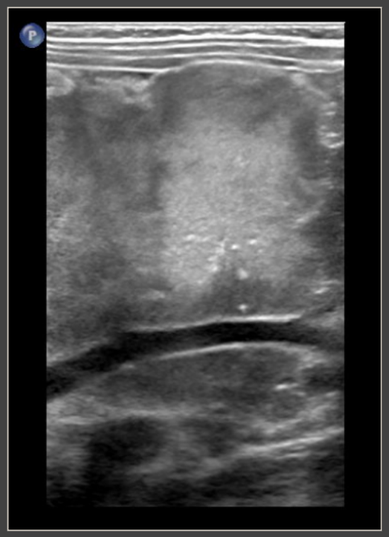


Lifting aorta

Neuroblastoma



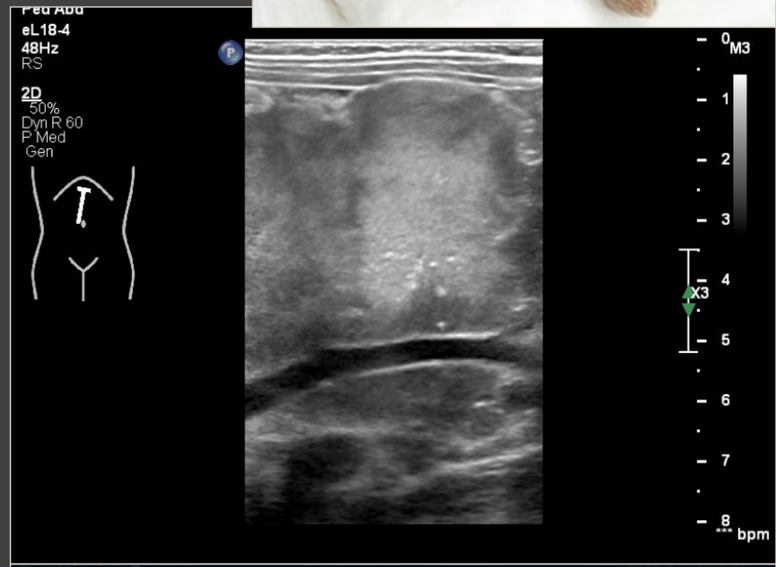


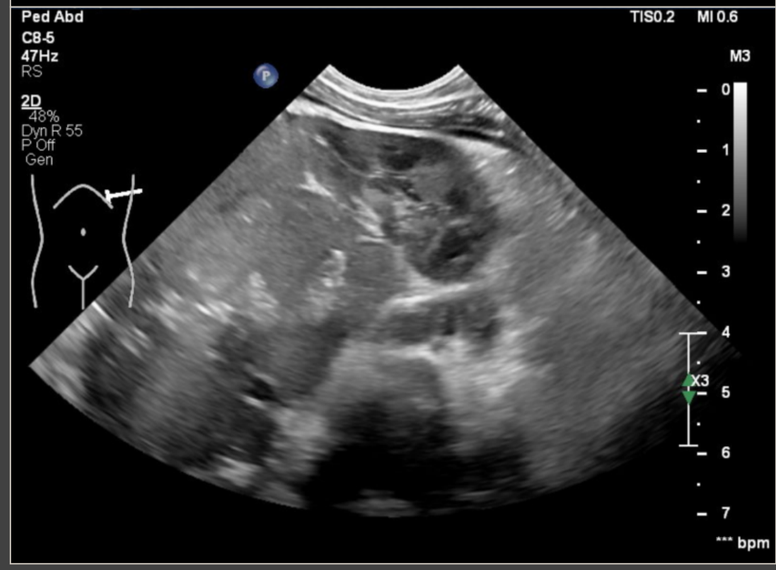


Neuroblastoma

- Most common extracranial solid tumor of childhood
- Embryonal tumor of sympathic nervous system
  - > 1/3 arises in the adrenal gland
- 90% of case before 5 years of age
- Can present antenatally
- Heterogenous group
  - Can show spontaneous regression in infants
  - Poor prognosis in high risk tumors with distant metastasis

Neuroblastoma





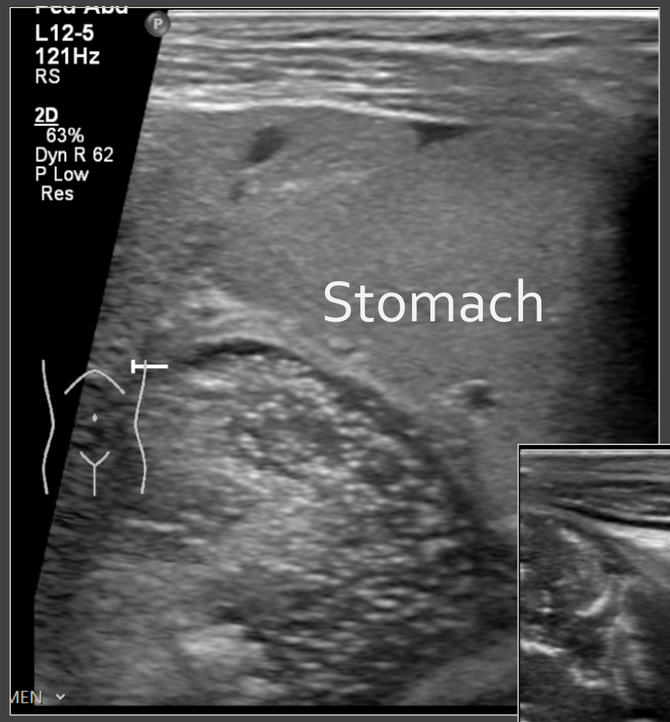
Discriminating imaging features:

- Growth pattern: encasing vessels/lifting aorta
- Internal calcifications
- Marked diffusion restriction

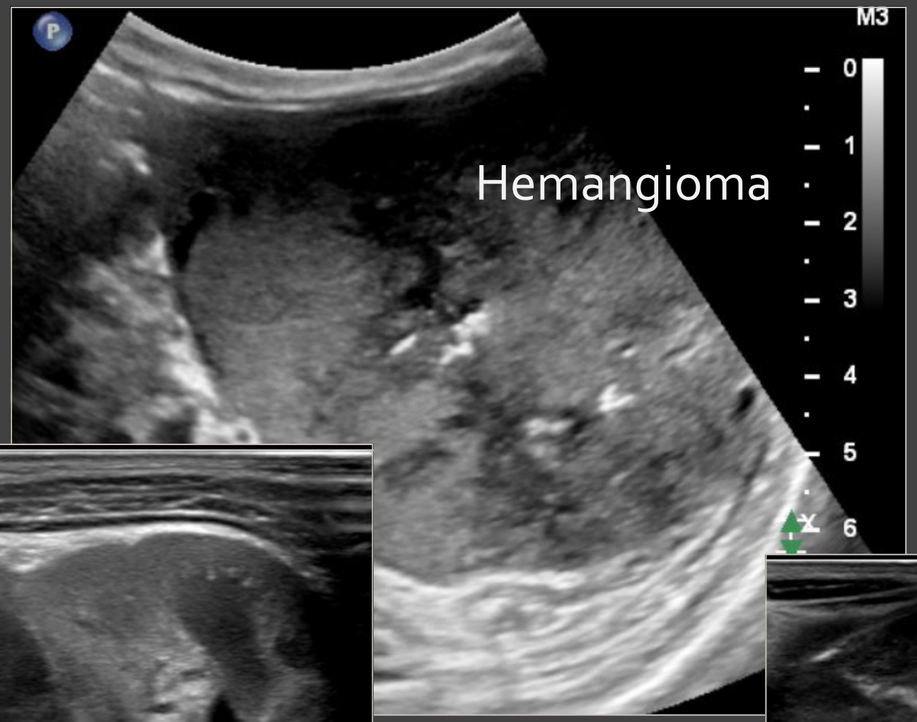
Tumor markers:

Urine metabolites (Catecholamines: HVA and VMA)

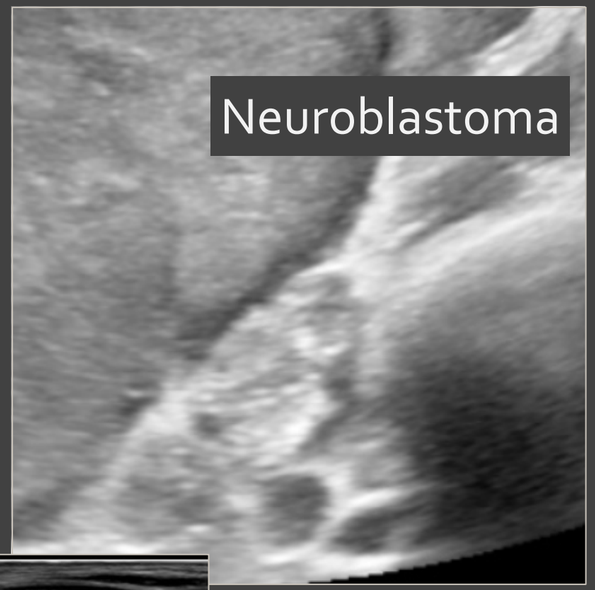
Internal calcifications



Stomach



Hemangioma


Neuroblastoma

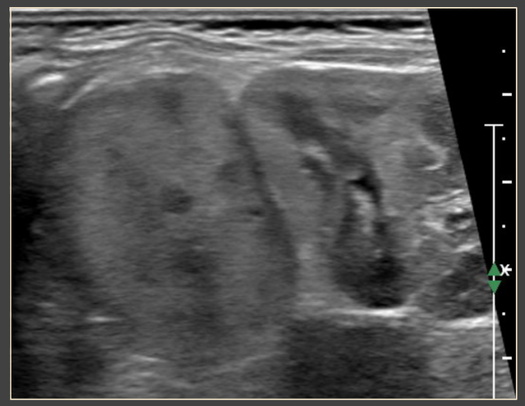

Neuroblastoma

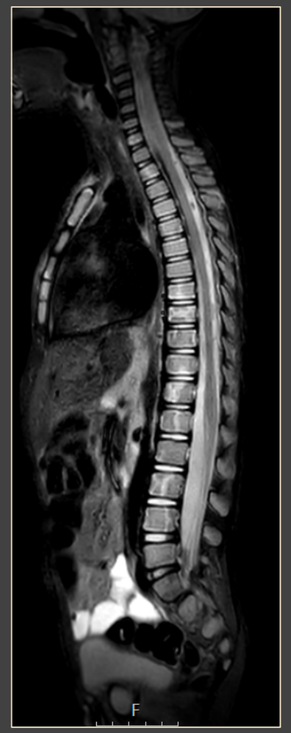

Neuroblastoma

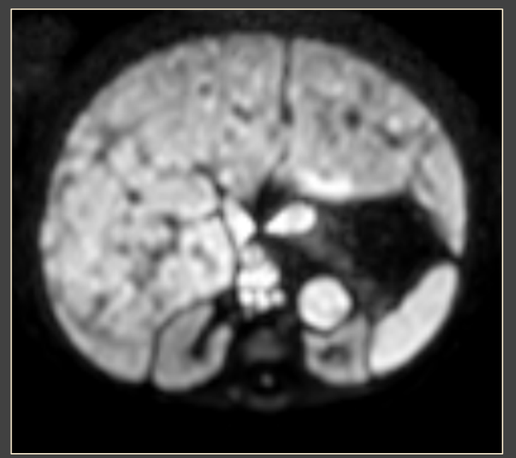
Clinical picture

3 main groups:

- Localized disease (40%)
- Disseminated disease (50%)
- 4S/MS disease (< 18 mo with mets to liver, skin or bone marrow)





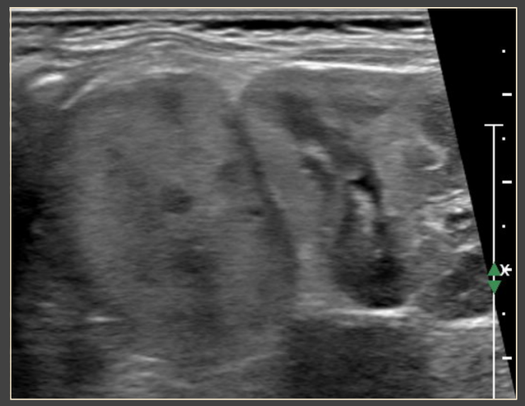


Stratification

- Low

- Intermediate

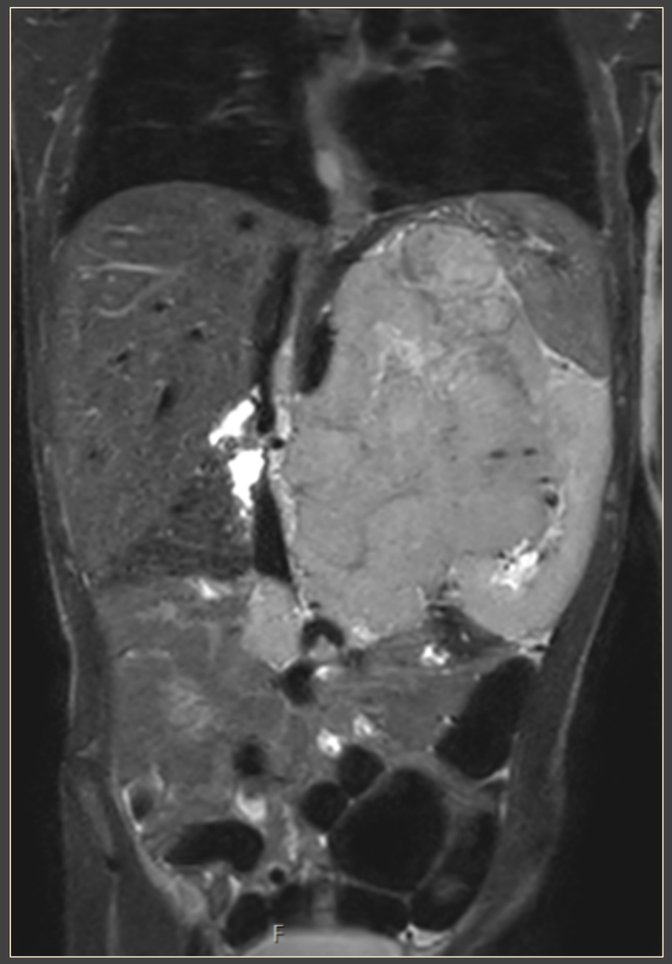
- High risk



Depending on: Age, stage and tumor biology (MYCN amplification)

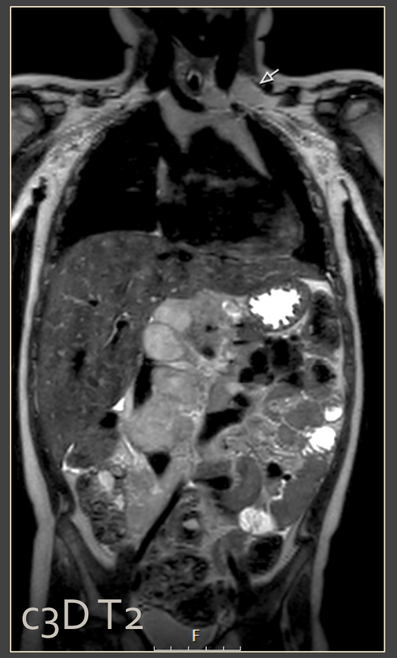
Imaging in Neuroblastoma

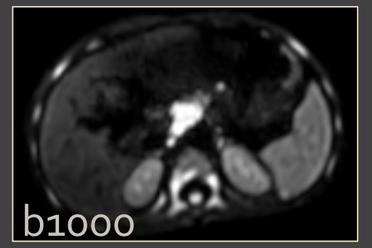
- Ultrasound
- MRI (or CT) for primary tumor
- [^{123}I] mIBG for local and distant staging

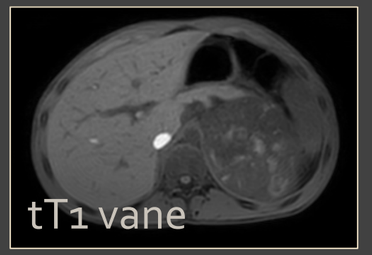


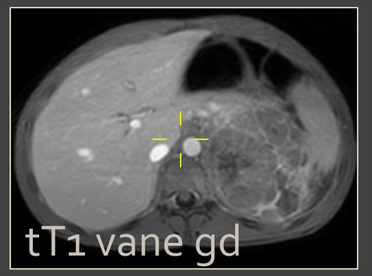
Our MRI protocol

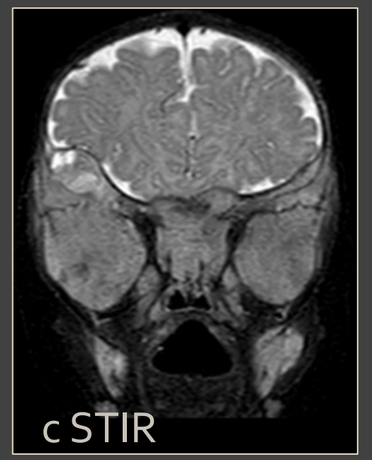
- c 3D T2
- t 3D t1 with fatsuppression before and after gd
- t DWI (b0,100 and 1000)
- Spine: s T1 TSE and s STIR
- Brain: c STIR, t T2, s T1, t DWI, 3D T1 gd



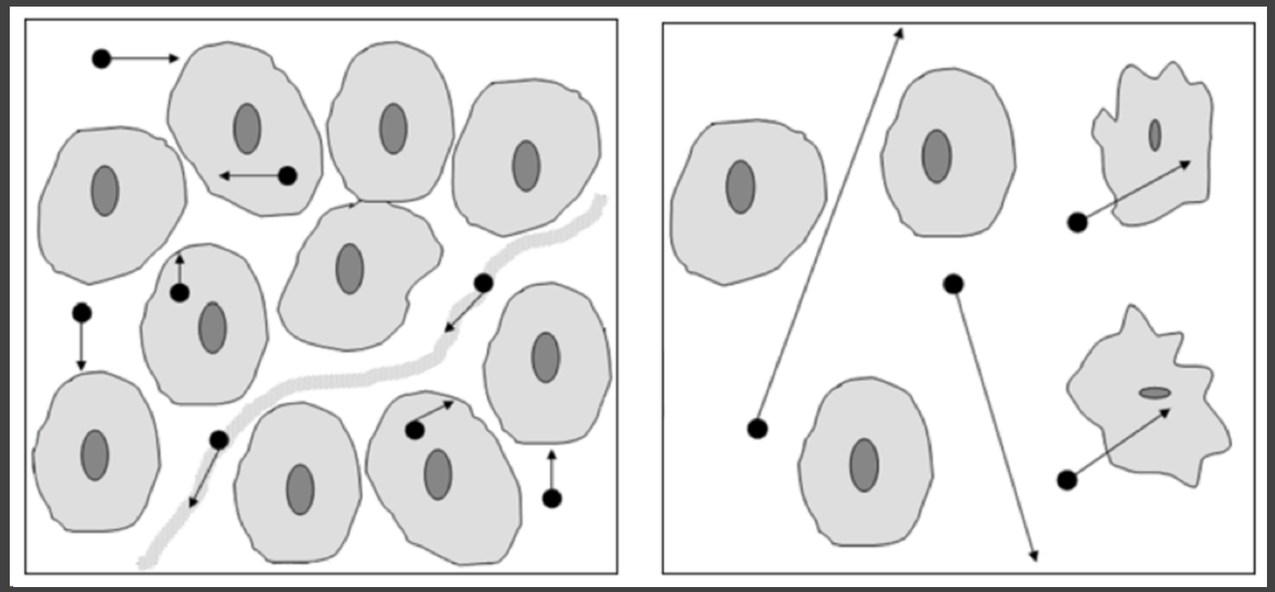






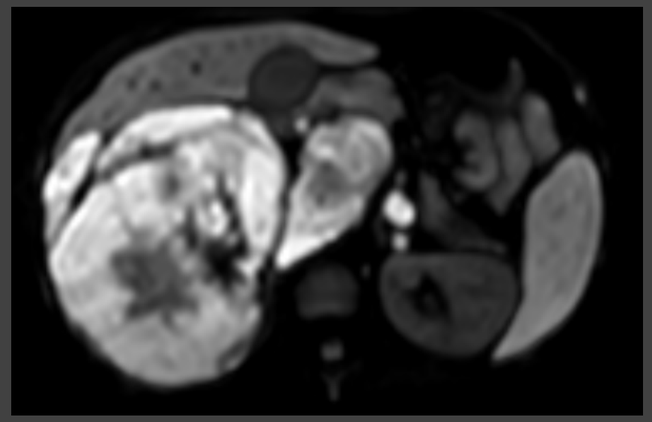


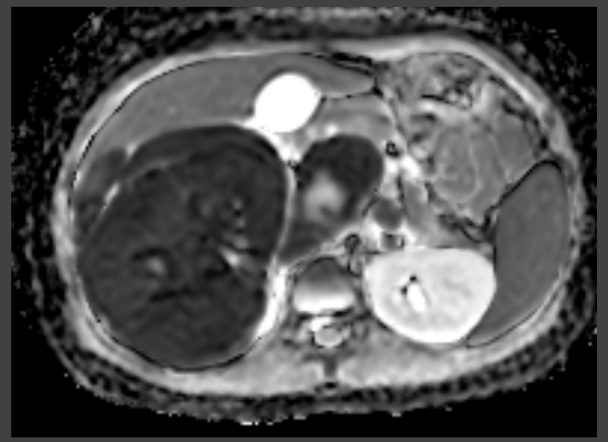
Diffusion Weighted Imaging (DWI)

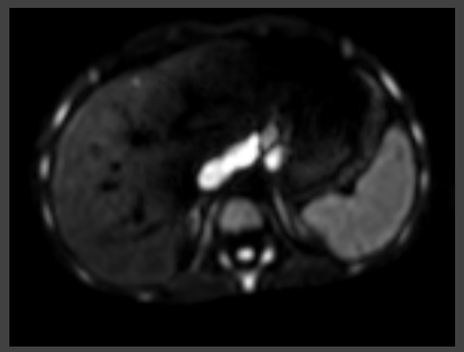


Role in Detection:

- Higher sensitivity
- Reading time







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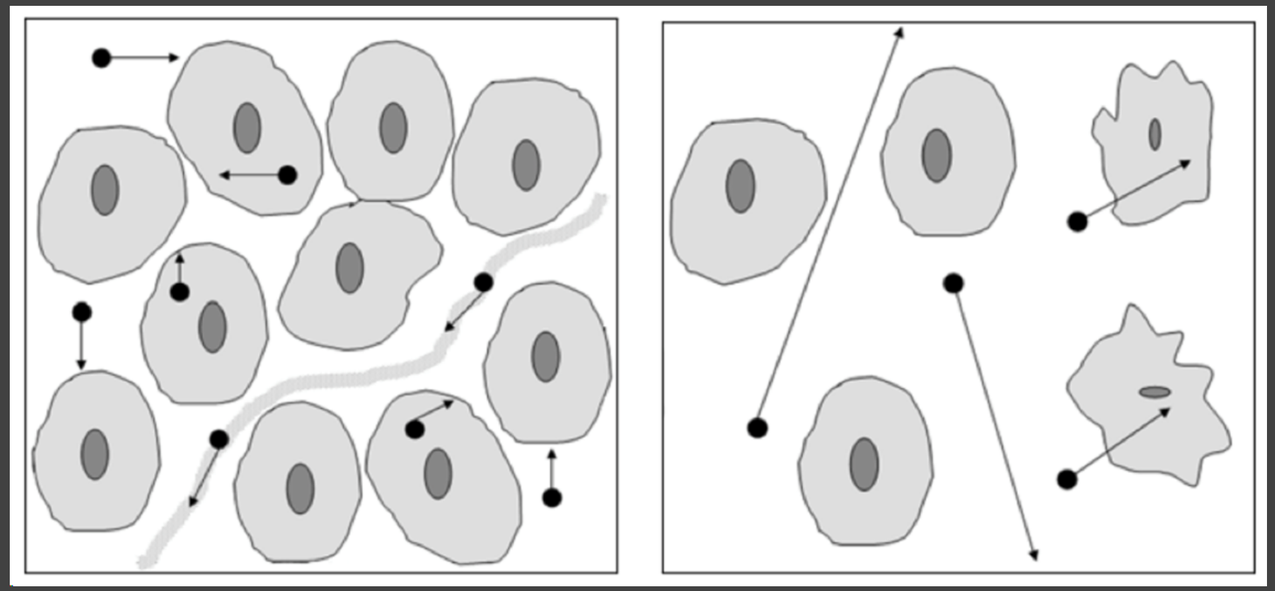


Role in Characterisation:

ADC Neuroblastoma = 0.8

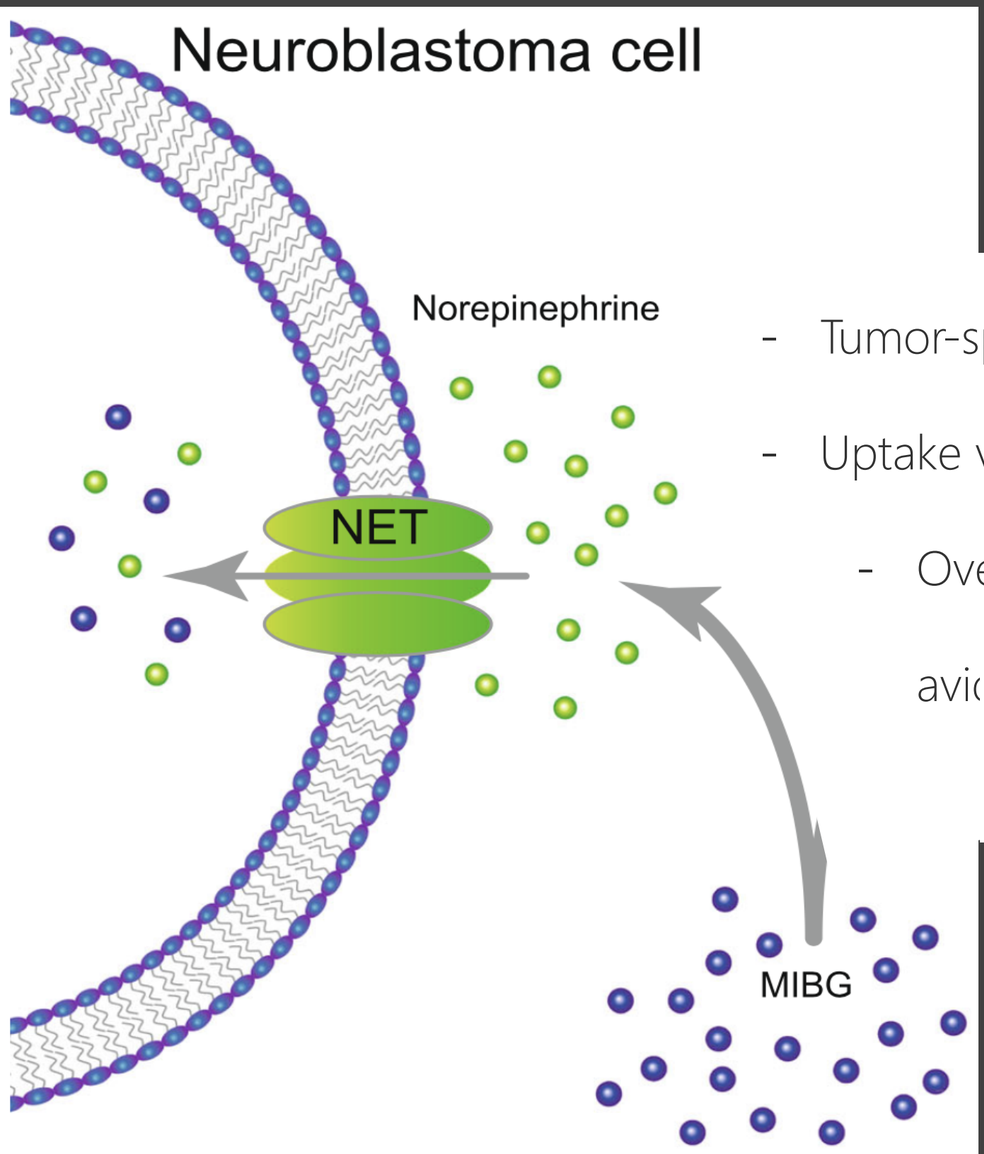
ADC Ganglioneuroma = 1.6

Diffusion-weighted MRI for differentiation of neuroblastoma and ganglioneuroblastoma/ganglioneuroma

Nina Gahr[a,*], Kassa Darge[b,1], Gabriele Hahn[c], Björn W. Kreher[d], Miriam von Buiren[e], Markus Uhl[f]

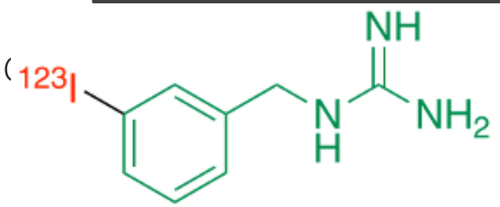
EJR 2011

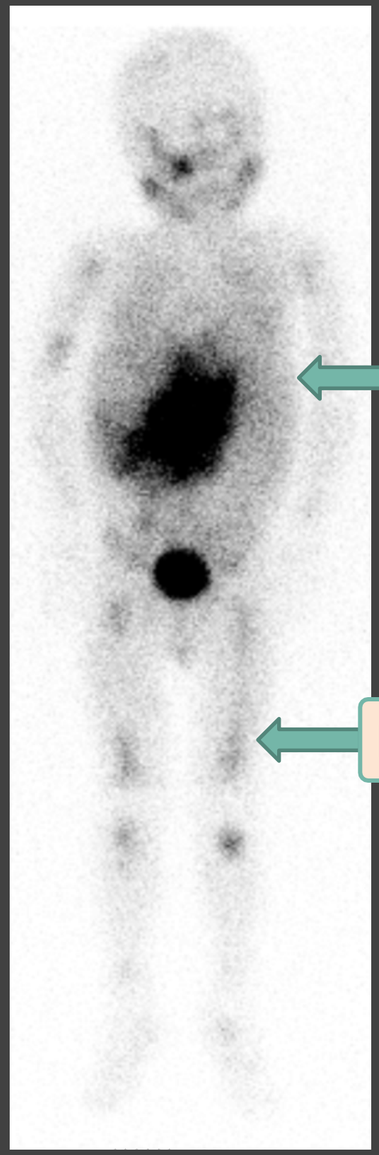
[^{123}I]mIBG



- Tumor-specific
- Uptake via norepinephrine transporter (NET)
  - Over 90% of neuroblastoma are MIBG-

avi

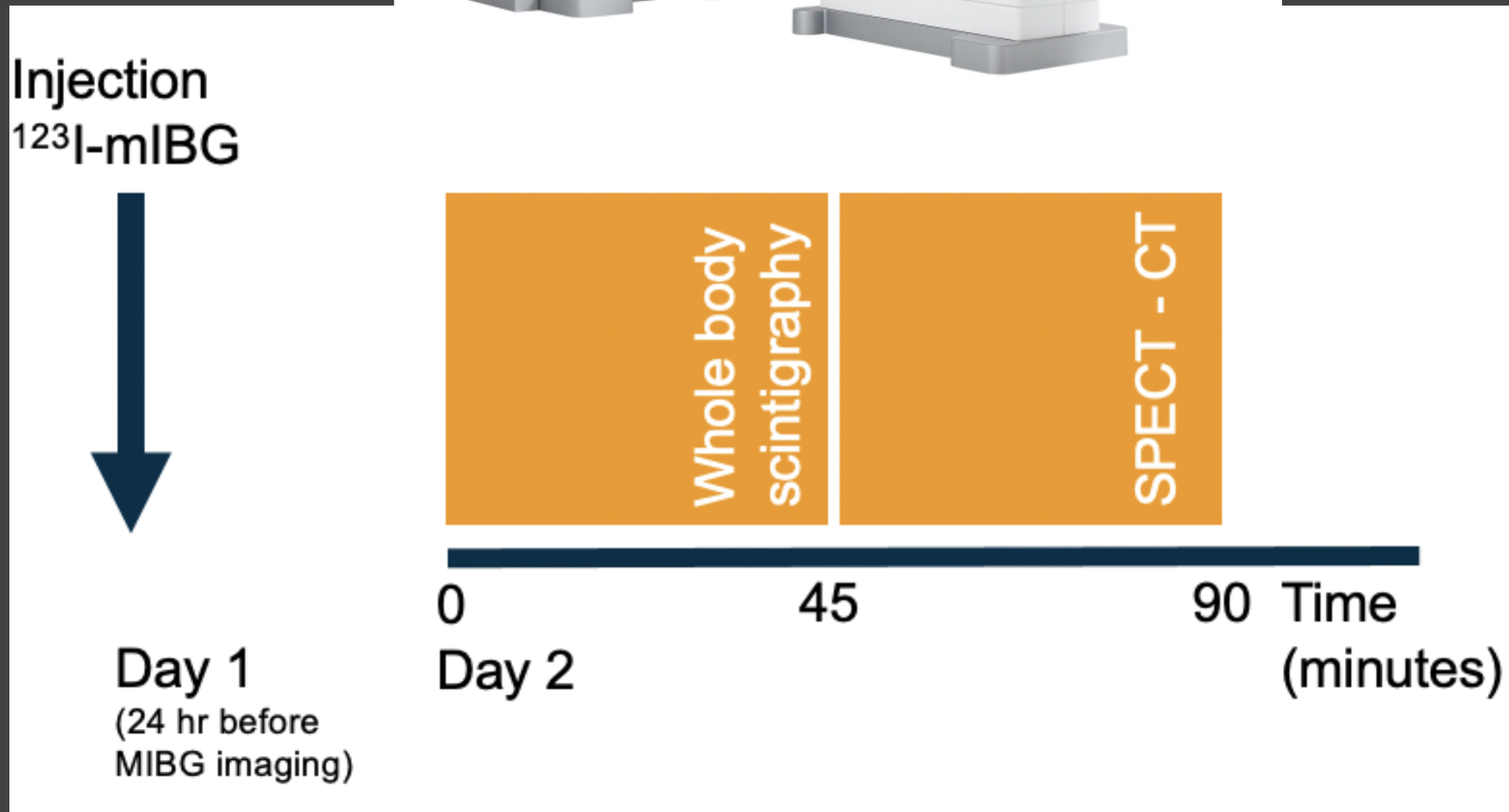




[^{123}I]mIBG scan

- **Scanning 24 hrs after injection of mIBG:**
- need for two visits

- **Total scan time ±1,5 hrs:**
- often need for sedation

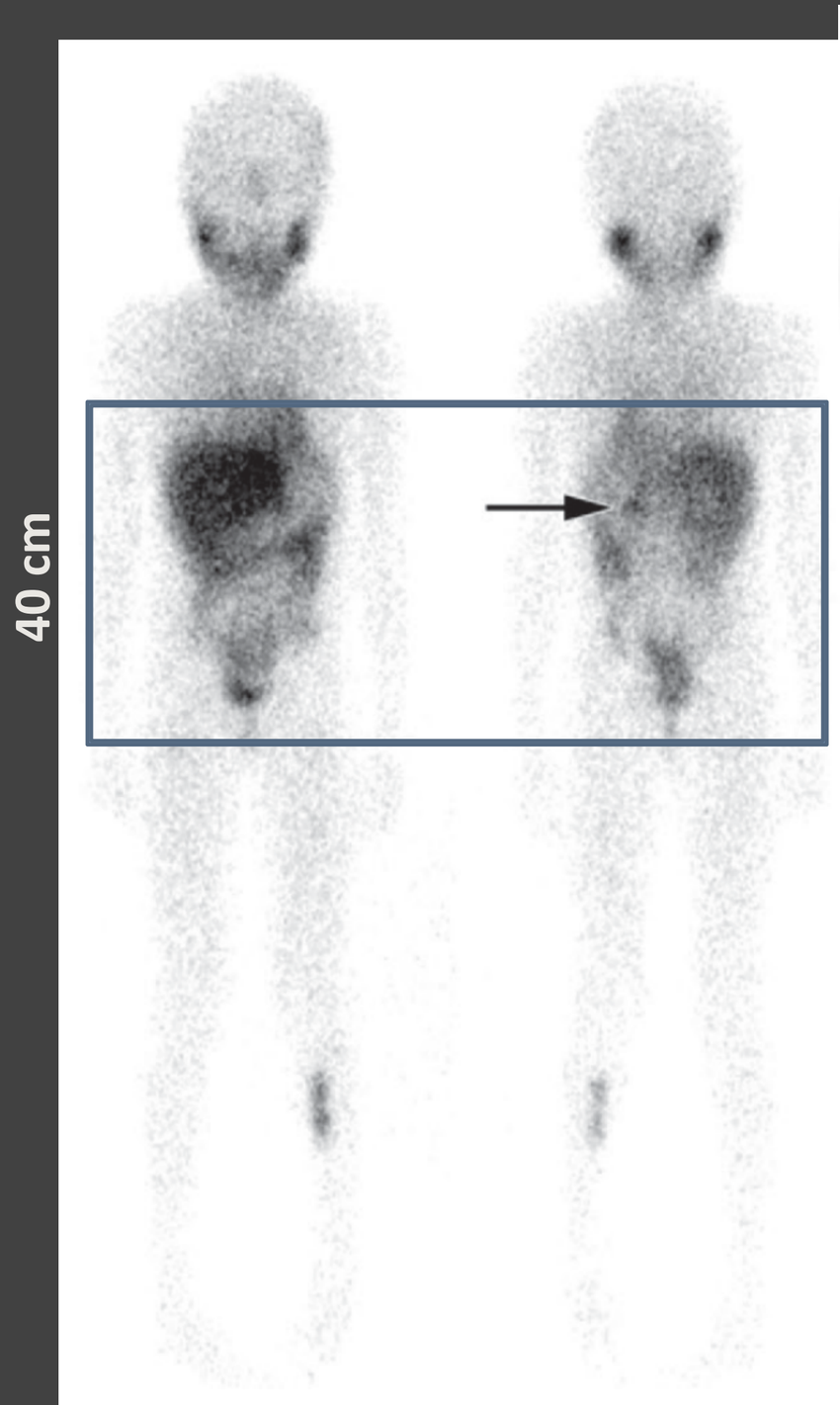


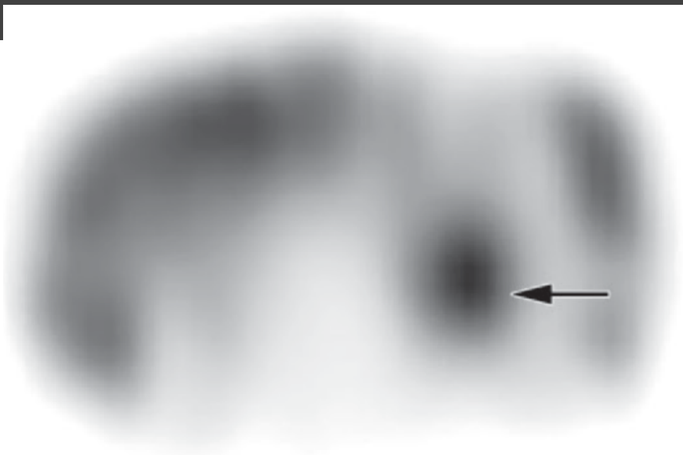
[^{123}I]mIBG scan

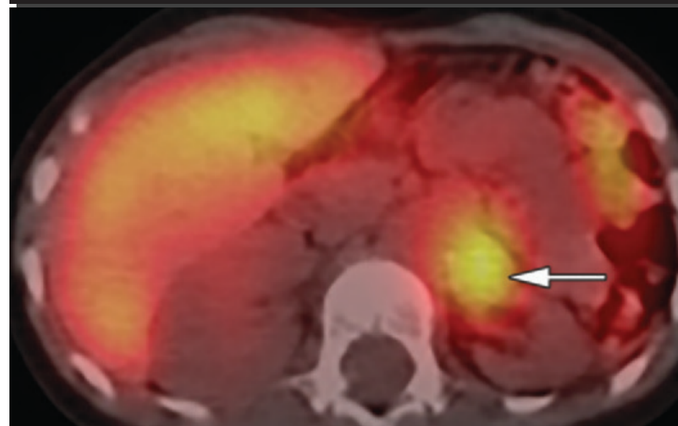
- Whole body scintigraphy (2D)
- SPECT-CT (3D): only 40cm range
- Limited resolution

- PET tracers:
  - [^{68}Ga]Ga-DOTA-peptides
  - [^{18}F]F-DOPA
  - [^{18}F]MFBG

Sharp 2016







SIOPEN mIBG skeletal score system

- Evaluates skeletal uptake on a 0-6 scale in 12 anatomical regions
- Max score 72

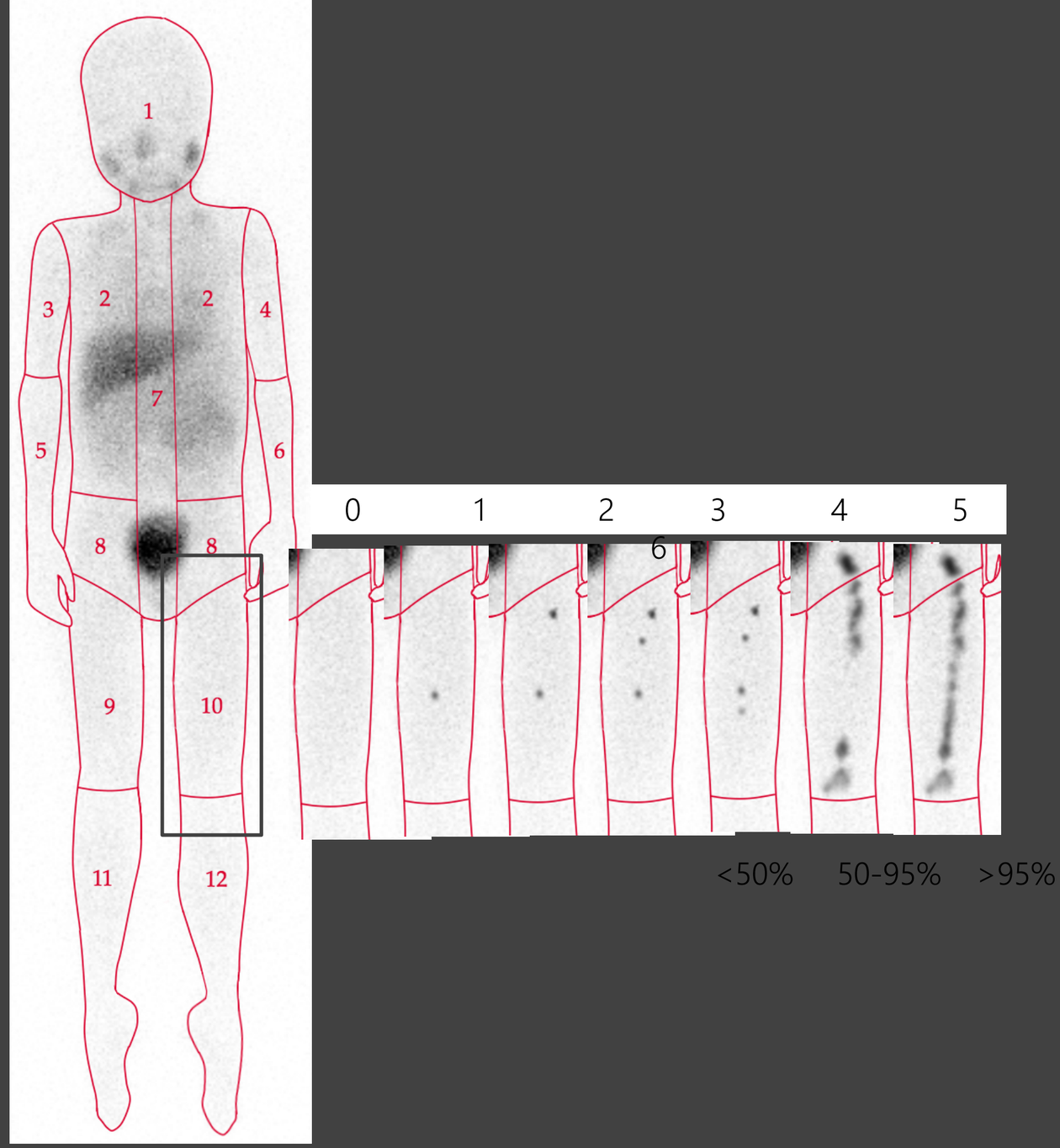
- At diagnosis: Increased risk with cumulative scores

- After induction: SIOPEN score >3 correlates with very poor outcome

[^{123}I]mIBG scan: SIOPEN score

- 12 skeletal segments

- End of induction total SIOPEN score > 3
- Bad prognosis > VERITAS



Content

Introduction

Imaging primary tumor - HR NBL 2 protocol

Distant metastases

Take home points

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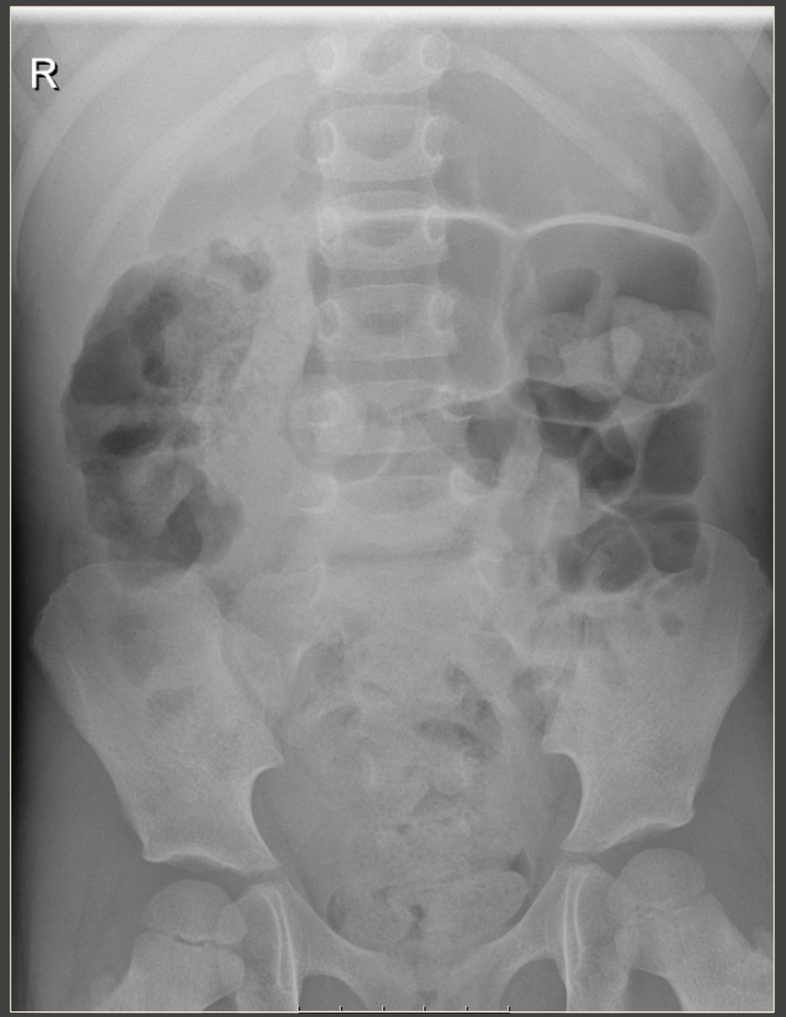
Distant metastases

Take home points

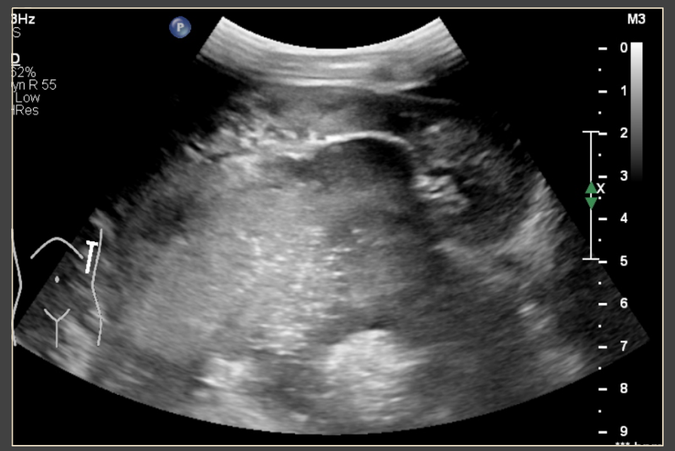
Case: 4-year-old girl with pain and fever

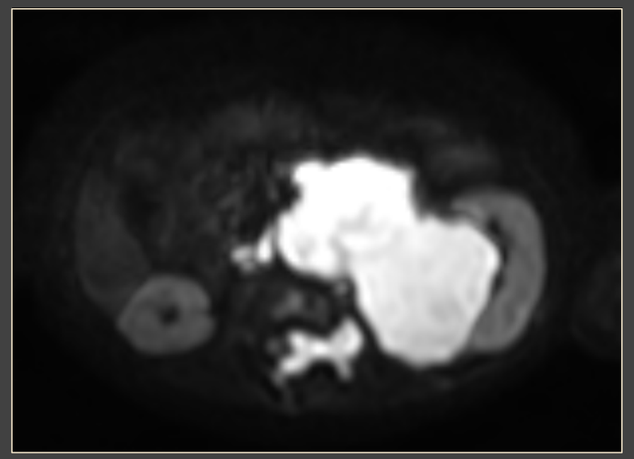


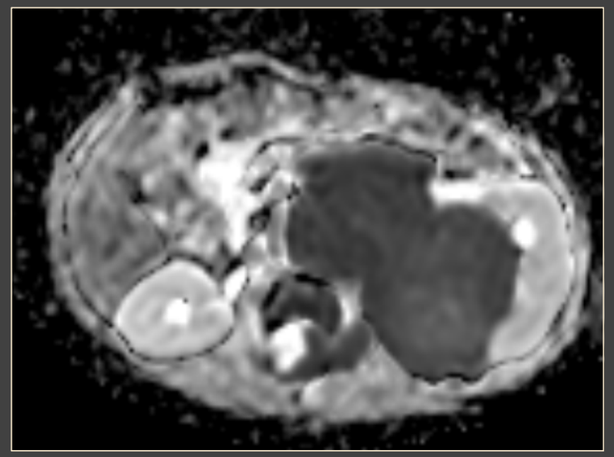
Febr 2019

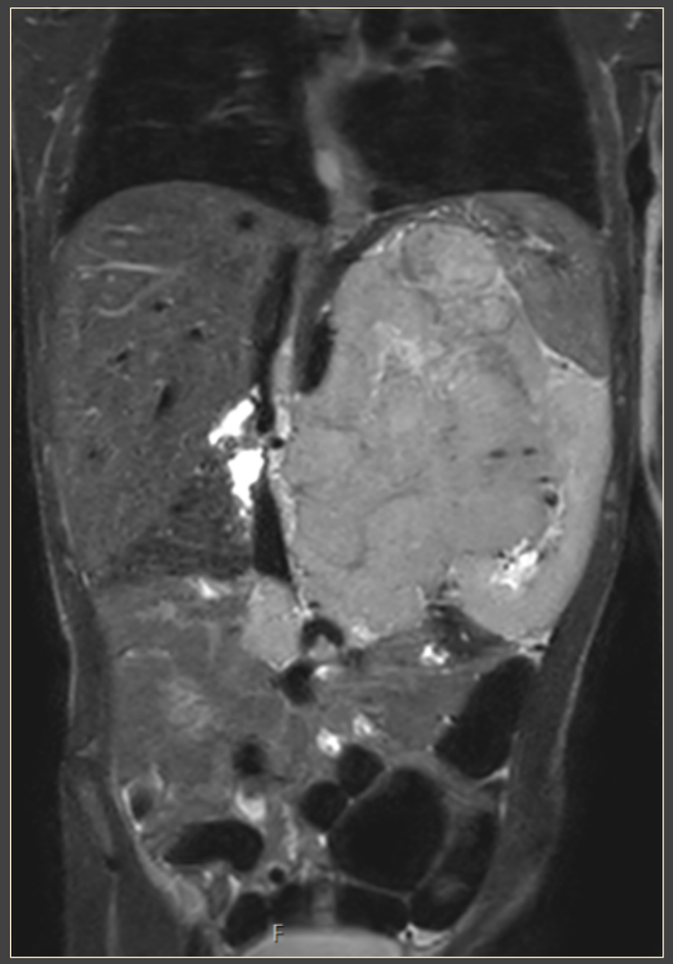
April 2019

Diagnosis: Neuroblastoma









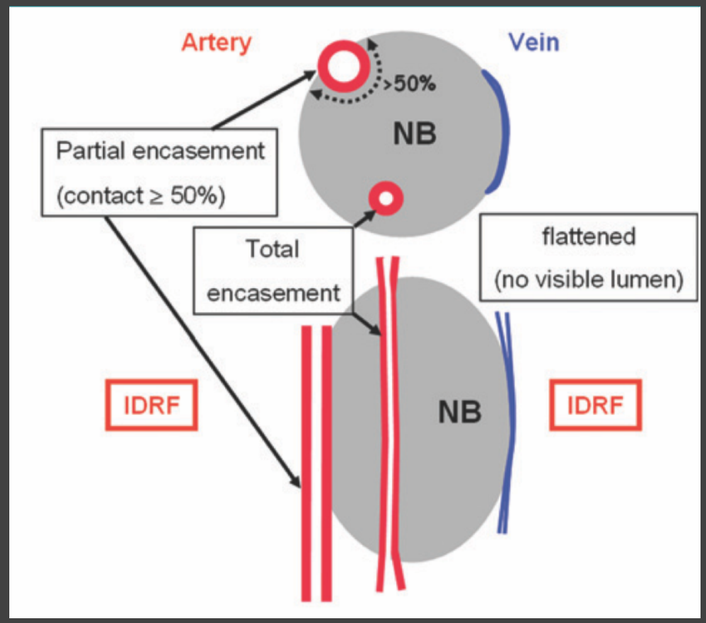
Staging

- INSS (International Neuroblastoma Staging System) - post-surgery
- INRGSS (International Neuroblastoma Risk Group)
  - Stage L1 – localised tumor, no IDRFs
  - Stage L2 – locoregional tumor, IDRFs +
  - Stage M – distant metastases
  - Stage MS – metastases < 18 mo, confined to skin, liver and BM

Image defined risk factors (IDRFs)

- Describe relationship of tumor with vital structures
  - Major vessels, airway, nerves
- Associated with high risk of surgical complications

- Warning for the surgeons
- Prognostic factor



Terminology

- Seperation (fatlayer visible)
- Contact (no fatlayer)
- Encasement: > 180 gr contact
- Compression (only for airways)
- Infiltration = involvement other than vessels

- According to Brisse et al 2011: renal vascular pedicle is IDRF postive when there is contact

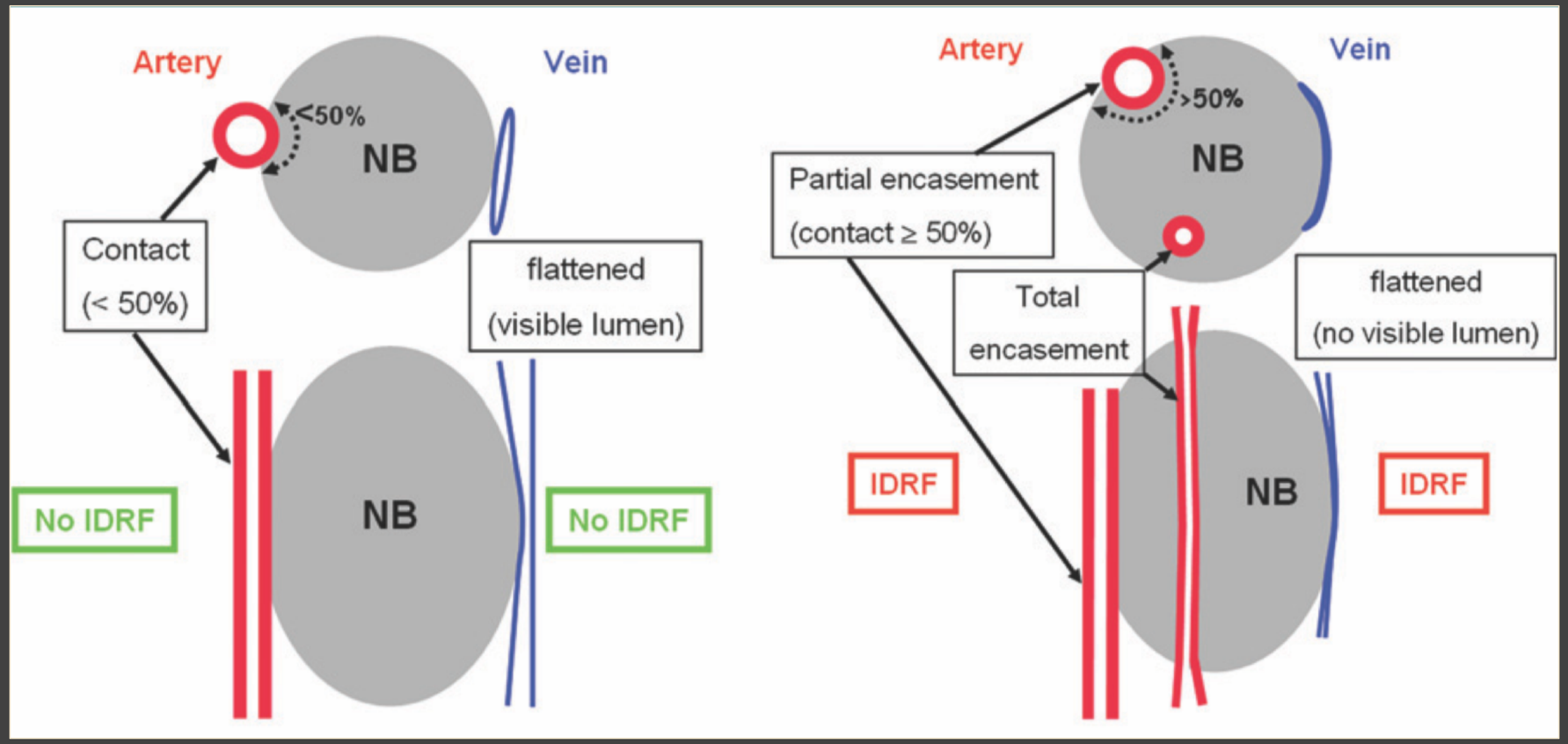
Major criteria

- Multicompartment tumor
- Encasement of large vessels
- Contiguous organ infiltration
- Significant spinal extention
- Airway compression

Image defined risk factors (IDRFs)

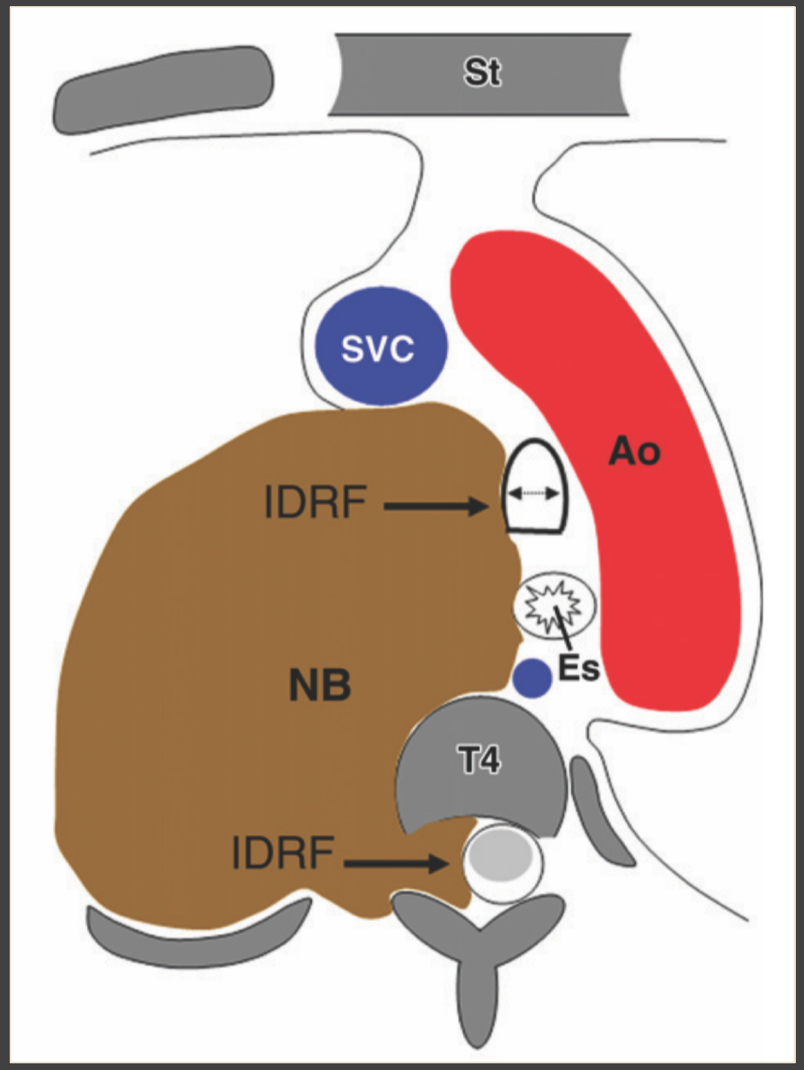
CONTACT

ENCASEMENT



* Adapted from" Guidelines for imaging and staging Neuroblastic tumors: Consensus report from the INRG project. Brisse HJ et al. Radiology 2011

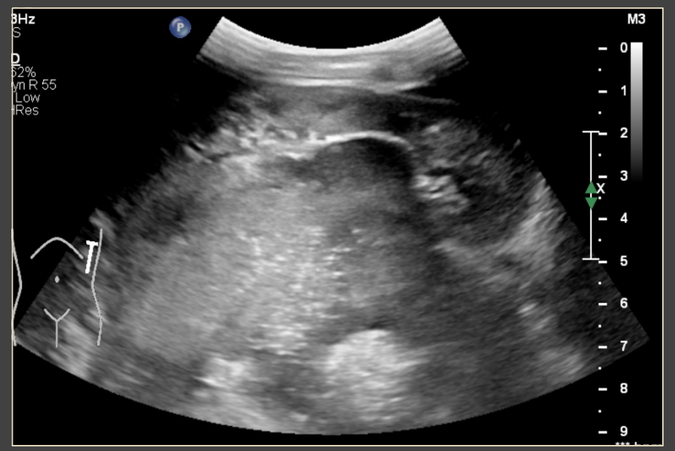
IDRFs

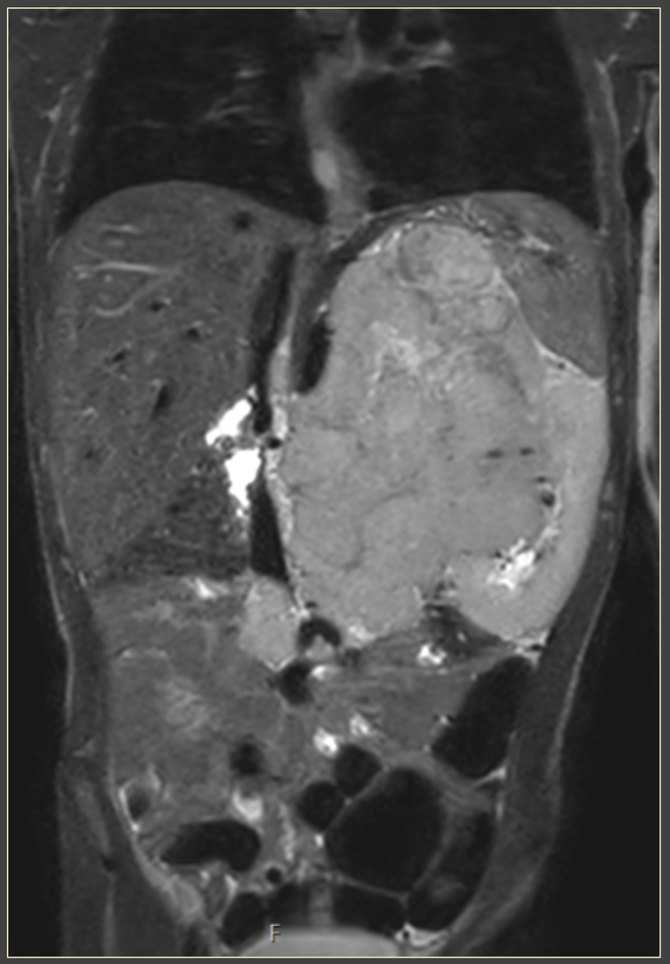


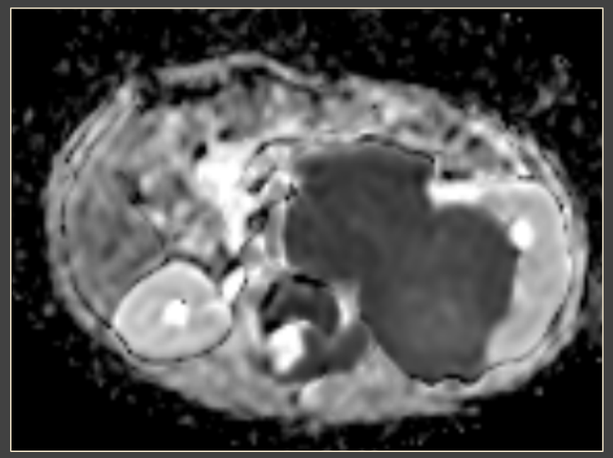
- IDRFs:
- 1/3 of the spinal canal is invaded
- Trachea diameter is compressed

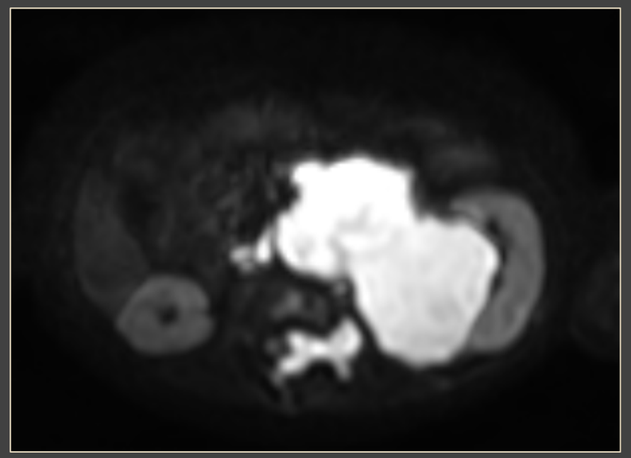
* Adapted from" Guidelines for imaging and staging Neuroblastic tumors: Consensus report from the INRG project. Brisse HJ et al. Radiology 2011

Case: 4-year-old girl with pain and fever









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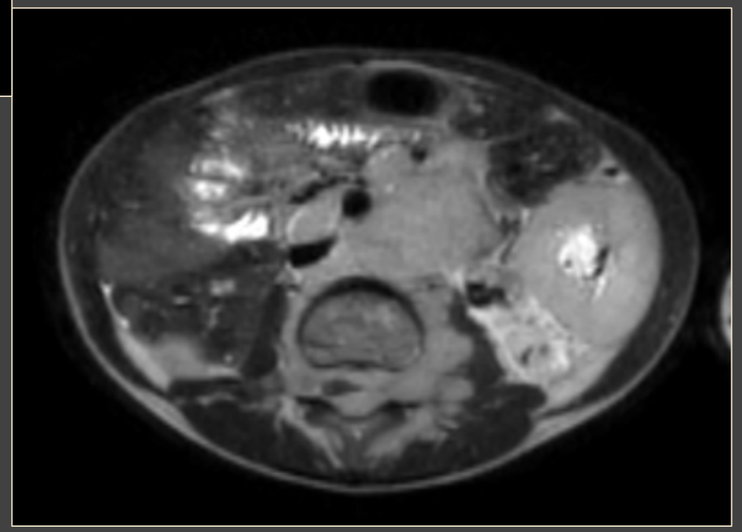
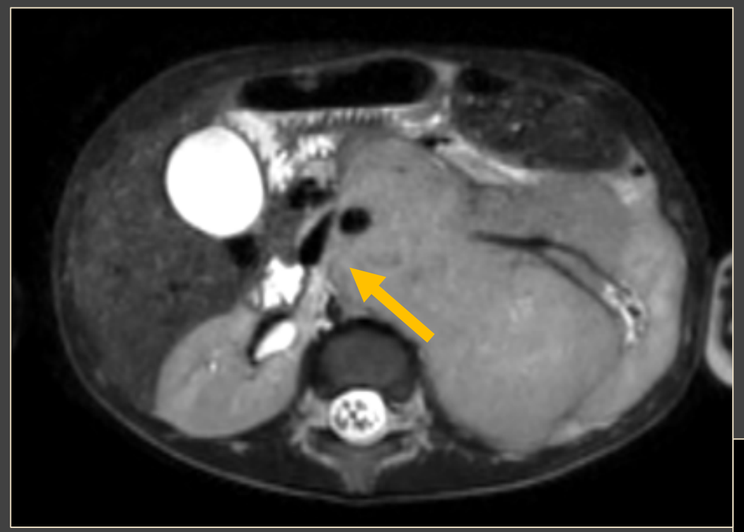
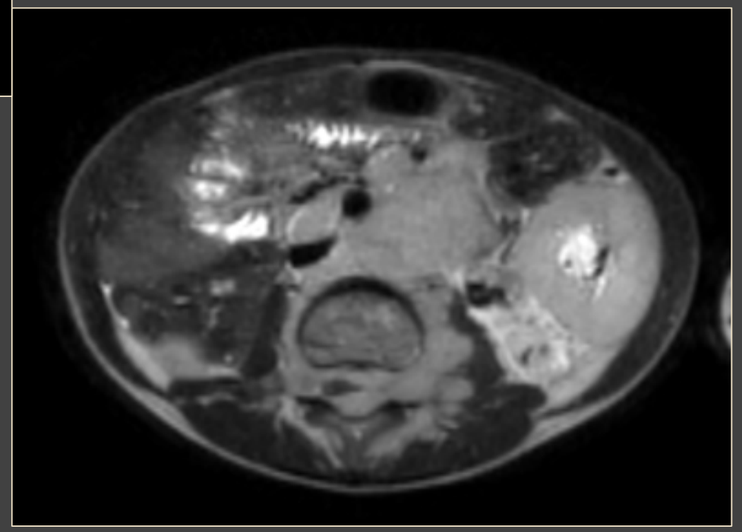
  - Stage MS – metastases < 18 mo, confined to skin, liver and BM

Staging – INRGSS Stage M – IDRFs?



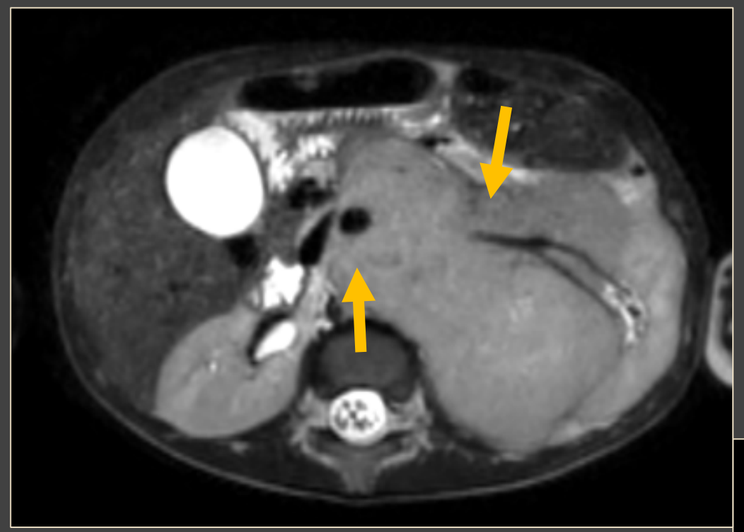
Staging – INRGSS Stage M – IDRFs?





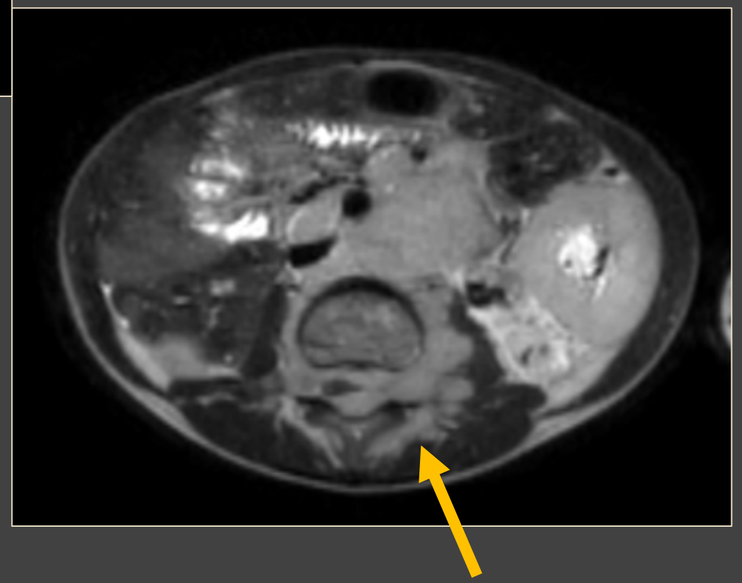
IDRF –
Contact with IVC, visible lumen

Staging – INRGSS Stage M – IDRFs?



IDRF –
Contact with IVC, visible lumen

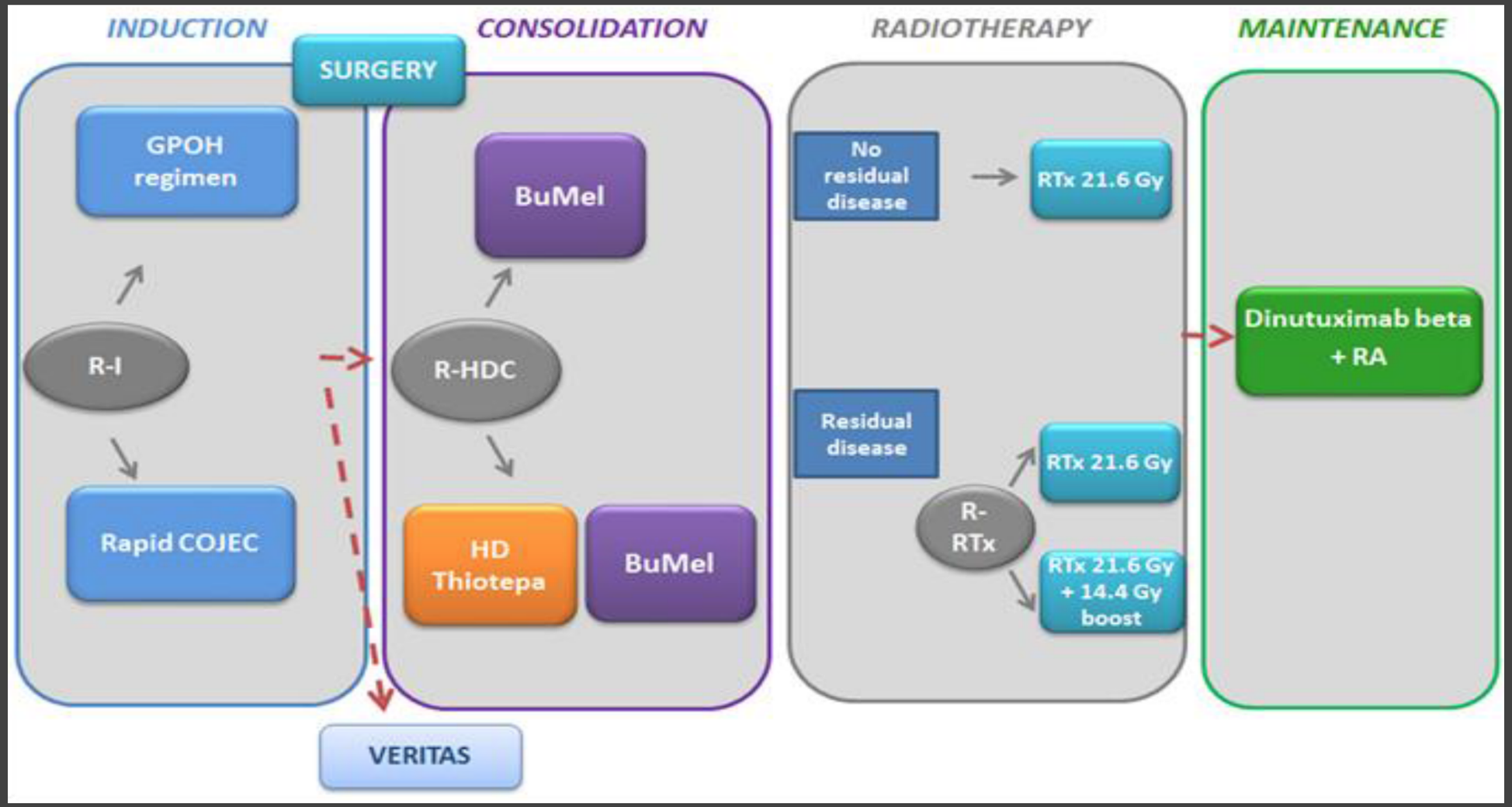
IDRF +
Encasement > 180: Aorta + renal vessels
Spinal canal extention > 1/3



High risk NBL

- Stage M over the age of 12 months, any MYCN status
- L2, M or Ms with MYCN amplification, any age
- 3-yrs EFS of 40%

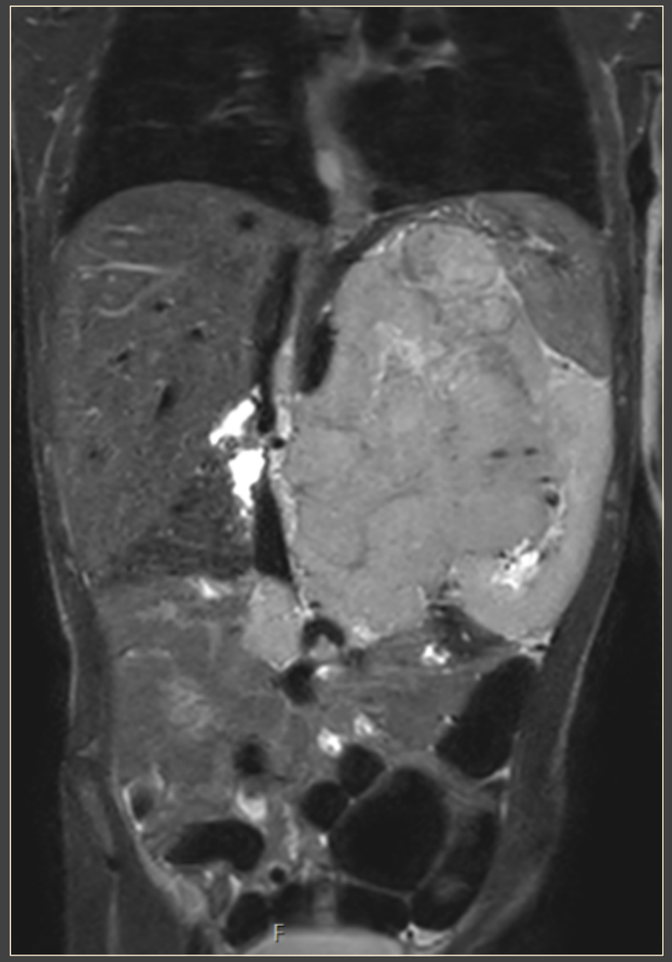
SIOPEN - HR NBL-2



Time points of imaging evaluation

- Staging
- Pre-operative assessment: CT or MRI?
- Identification of residual disease before RT: diagnostic criteria?
- Evaluation before maintenance
- Surveillance – relapse?

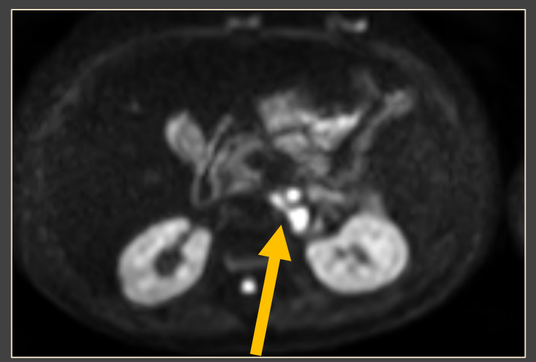
After extensive chemo: for surgical planning

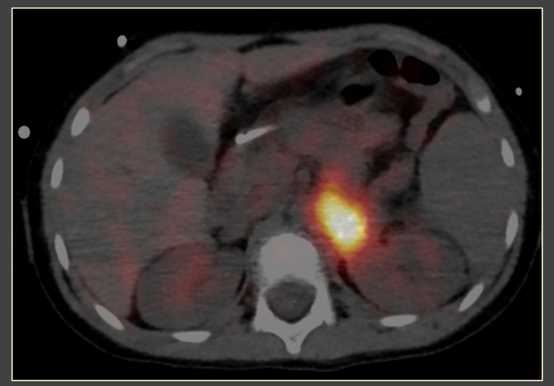


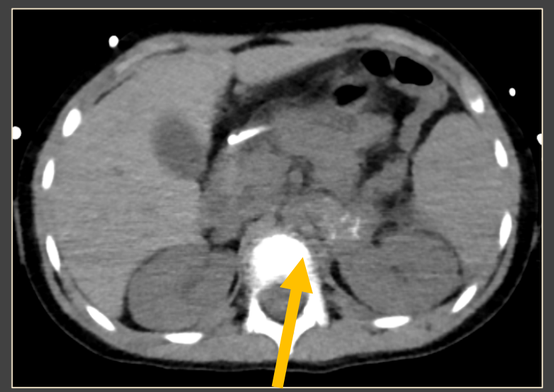
Staging

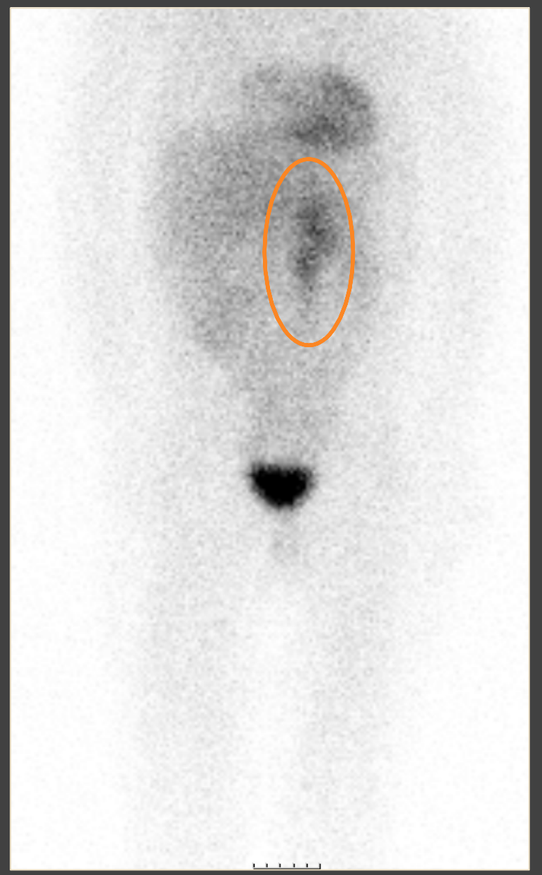
After chemo

After extensive chemo: for surgical planning

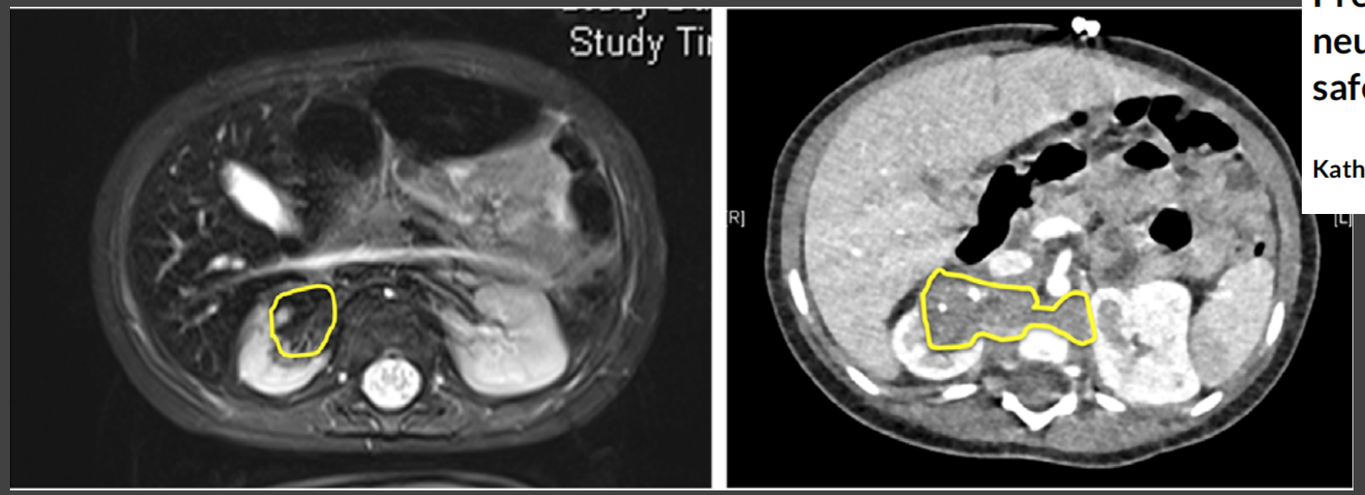








For surgical planning ~ CT > MRI



Received: 28 February 2019 | Revised: 7 July 2019 | Accepted: 10 July 2019

DOI: 10.1002/pbc.27955

Pediatric Blood & Cancer

RESEARCH ARTICLE

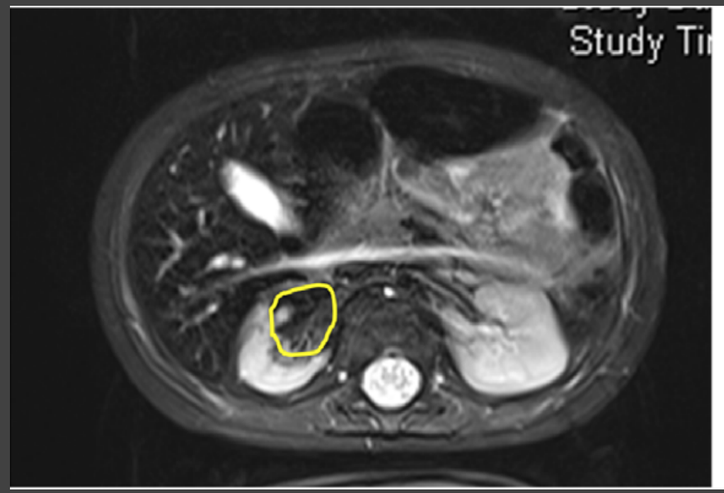
Preoperative computed tomography scanning for abdominal neuroblastomas is superior to magnetic resonance imaging for safe surgical planning

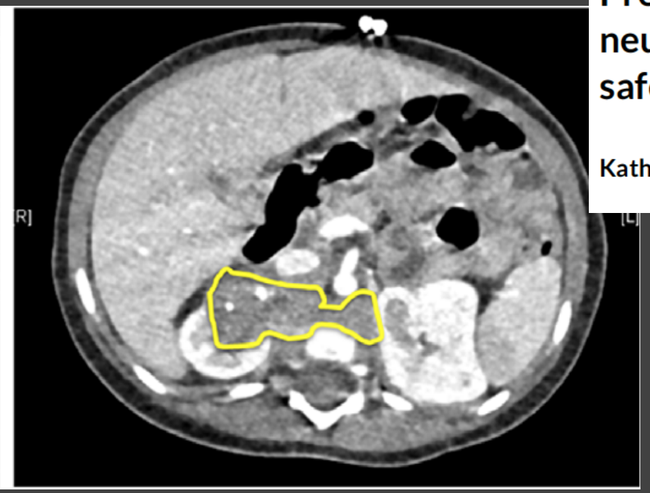
Katherine Burnand[1] | Giuseppe Barone[2] | Kieran McHugh[3] | Kate Cross[1]

N=24, surgery in 17 cases

Conclusion: MRI underestimated the extent of the disease in half of our patients considered for NBL resection. This may be due in part to tumor fibrosis, calcification, and chemotherapy. Preoperative CT scan is the best imaging modality to identify all IDRFs after chemotherapy to ensure safe surgery.

For surgical planning ~ CT > MRI





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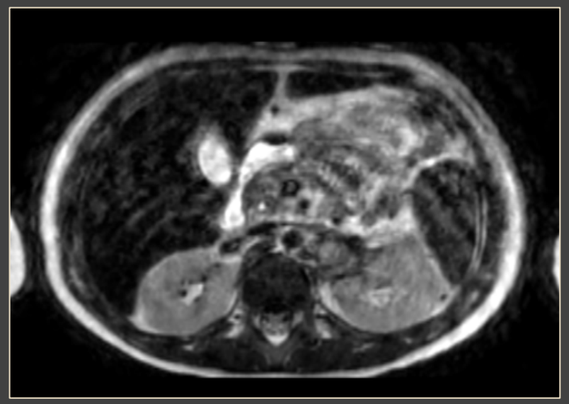

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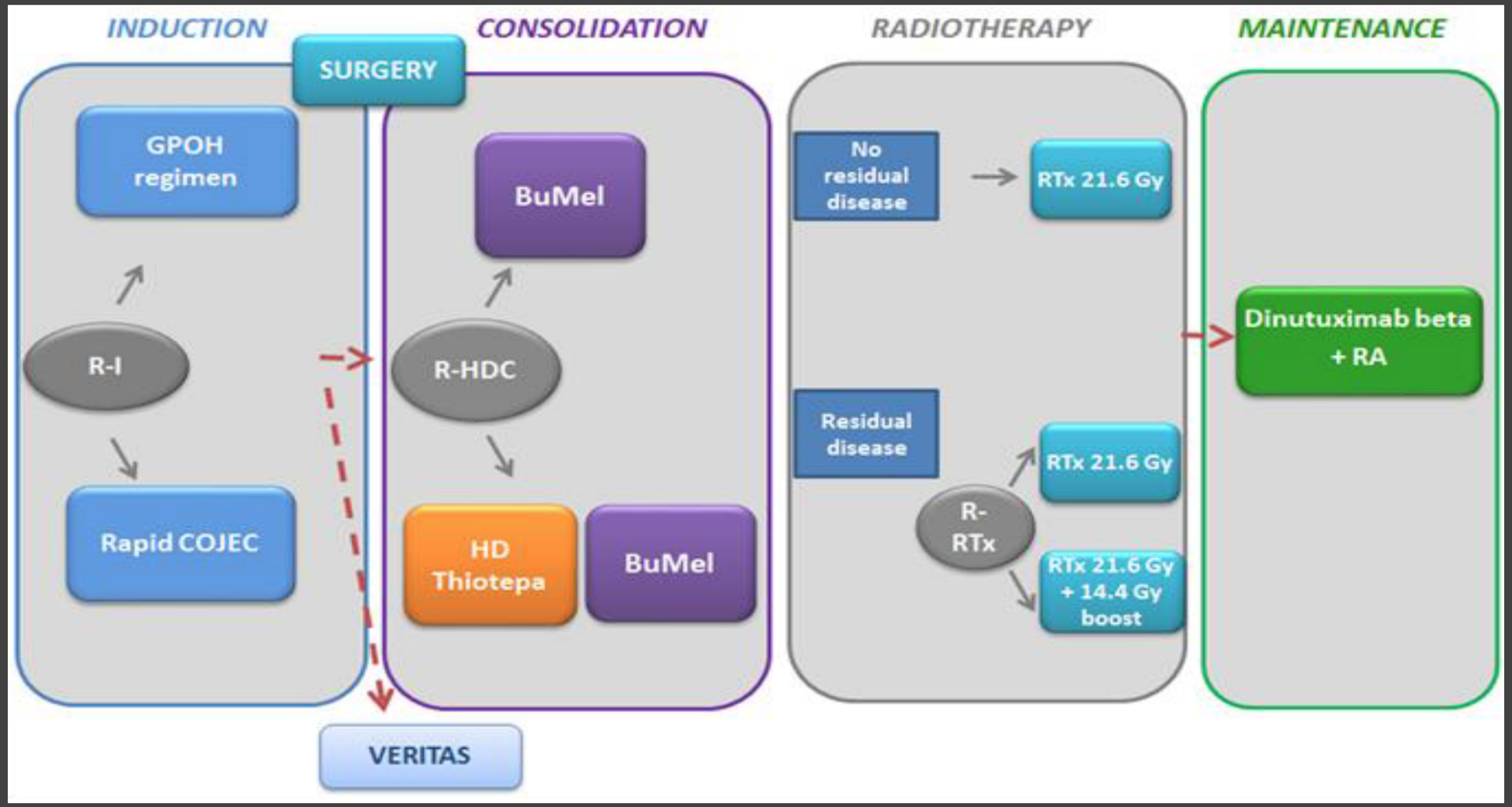
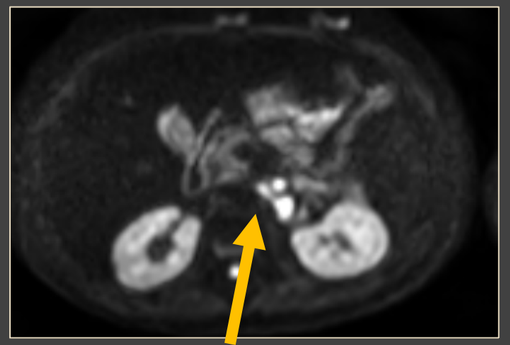
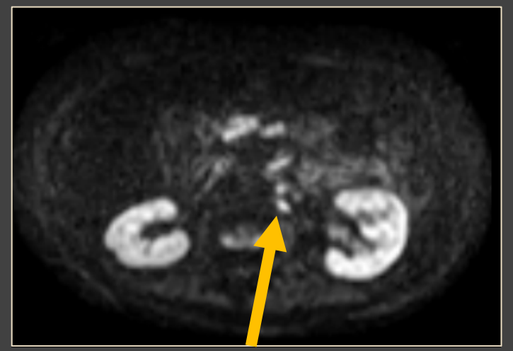
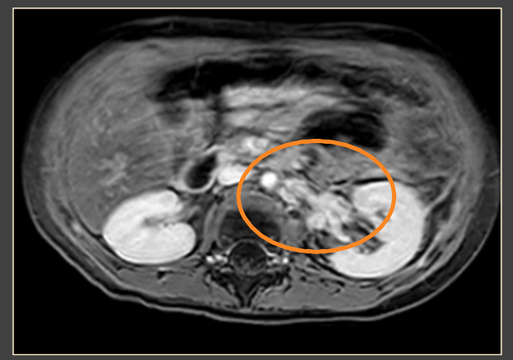
SIOPEN - HR NBL-2



After resection. Pre-RT assessment





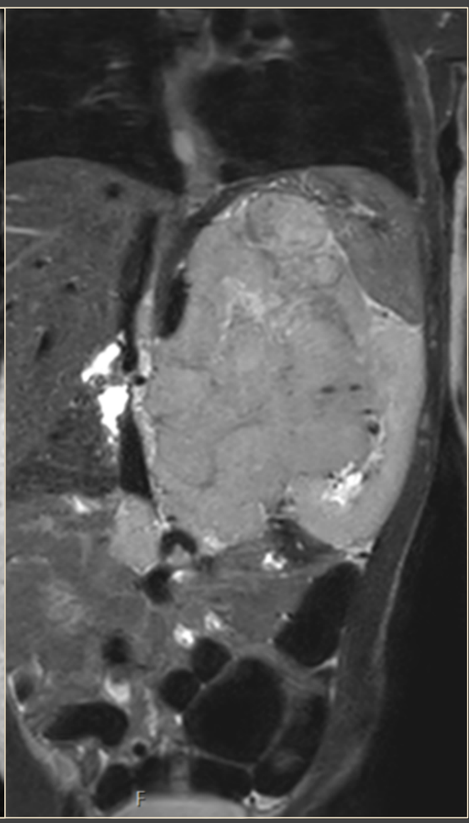


Before surgery

After surgery

After surgery

Before surgery



Staging

Definition of residual disease

- MRI shows definite residual tumor and/or
- mIBG shows residual tumor and/or
- Surgical report mentions residual tumor (marks with MRI compartible clips)

Definite residual disease?

SIZE?

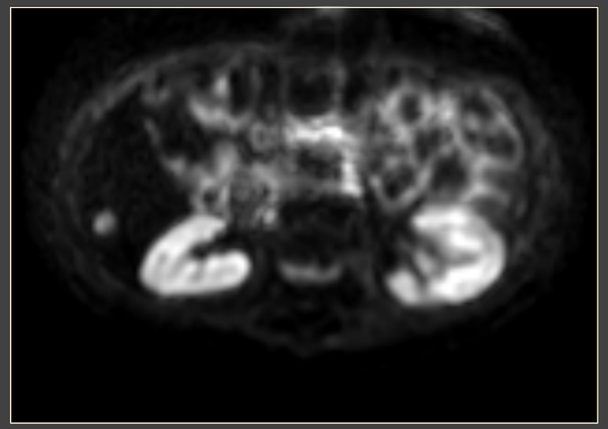
DWI?

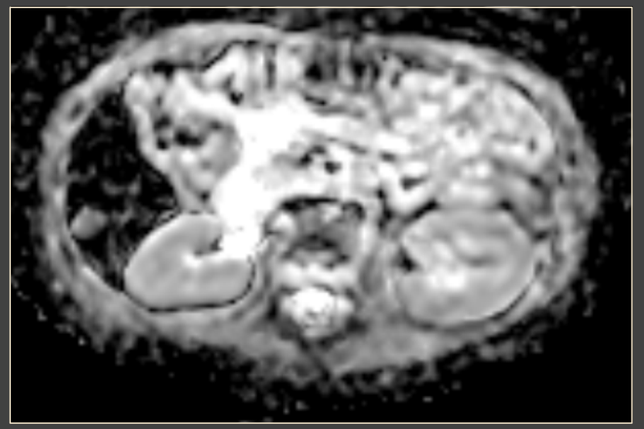
MIBG?

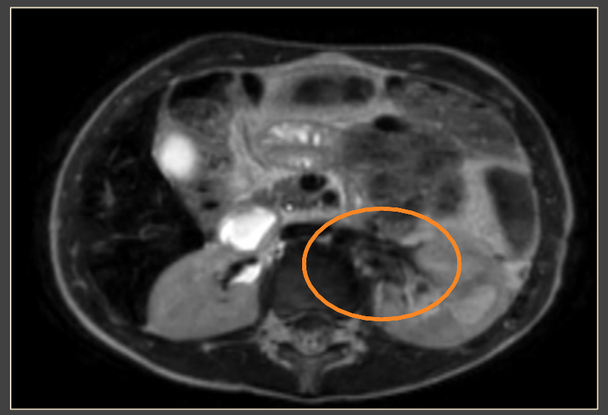
Definitive residual disease?

- Residual disease:
  - All solid tissue that was present pre-surgery and is still visible should be considerd positive
  - Use previous imaging and surgical report!
  - Fibrosis/post-operative changes versus viable tumor?
    - MIBG is not very sensitive for small lesions
    - Role for DWI or 48-hours post-resection MRI ?

Currently in CR?

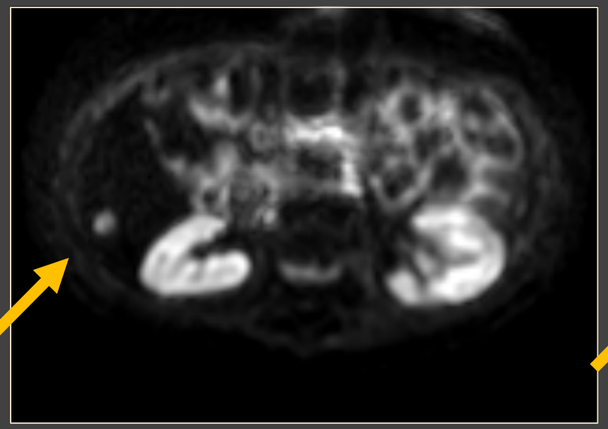


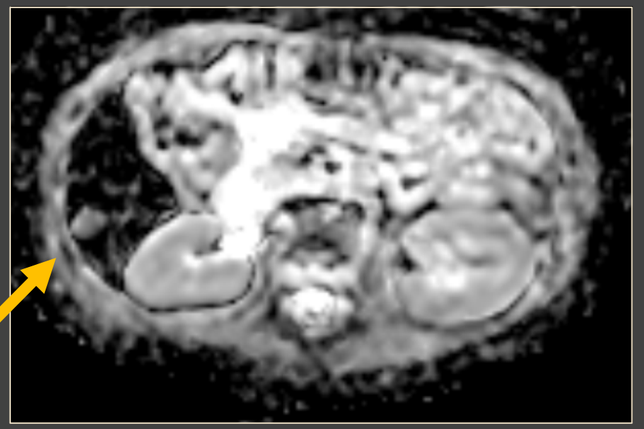


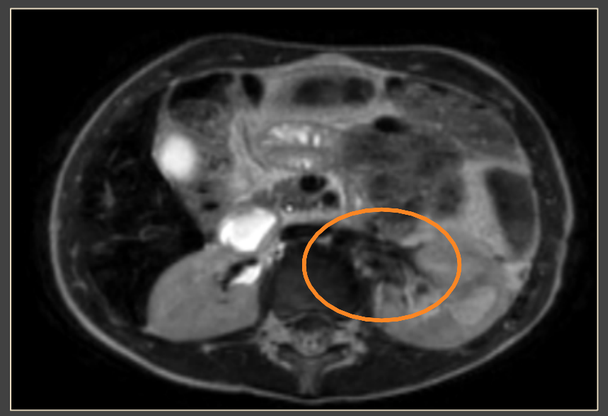


ADC value 1.1

Currently – CR?



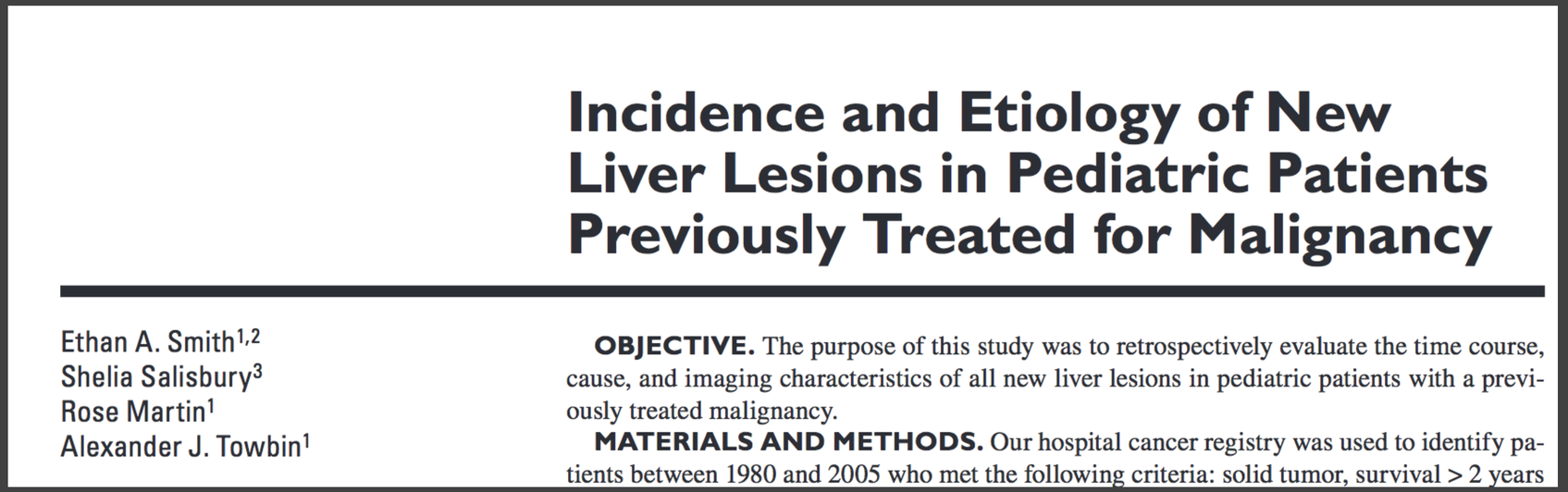




ADC value 1.1

FNH like lesion

FNH like lesions



Incidence and Etiology of New Liver Lesions in Pediatric Patients Previously Treated for Malignancy

Ethan A. Smith[1,2]
Shelia Salisbury[3]
Rose Martin[1]
Alexander J. Towbin[1]

OBJECTIVE. The purpose of this study was to retrospectively evaluate the time course, cause, and imaging characteristics of all new liver lesions in pediatric patients with a previously treated malignancy.

MATERIALS AND METHODS. Our hospital cancer registry was used to identify patients between 1980 and 2005 who met the following criteria: solid tumor, survival > 2 years

- N=273
- 3% livermets
- 14% benign liverlesions

FNH like lesions

- Related to vascular injury
  - Alkylating agents (eg busulfan/ melphalan ~ BuMel)
  - Veno-occlusive disease
  - Radiotherapy
  - >>> not rarely after HR NBL treatment!

Content

Introduction

Imaging primary tumor - HR NBL 2 protocol

Distant metastases

Take home points

Content

Introduction

Imaging primary tumor - HR NBL 2 protocol

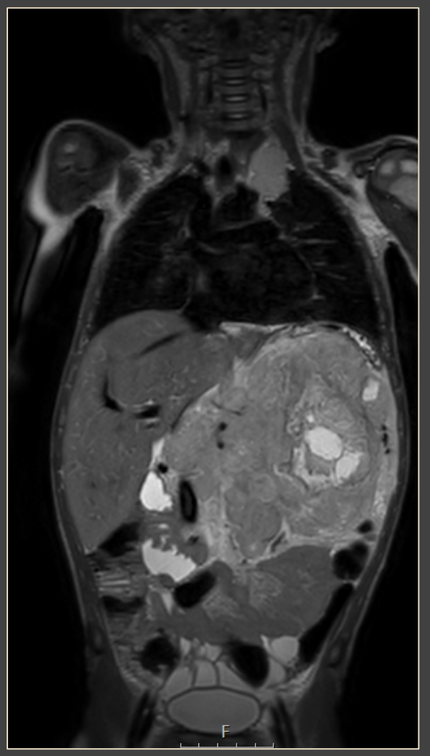
Distant metastases

Take home points

Role of MRI in distant metastases?

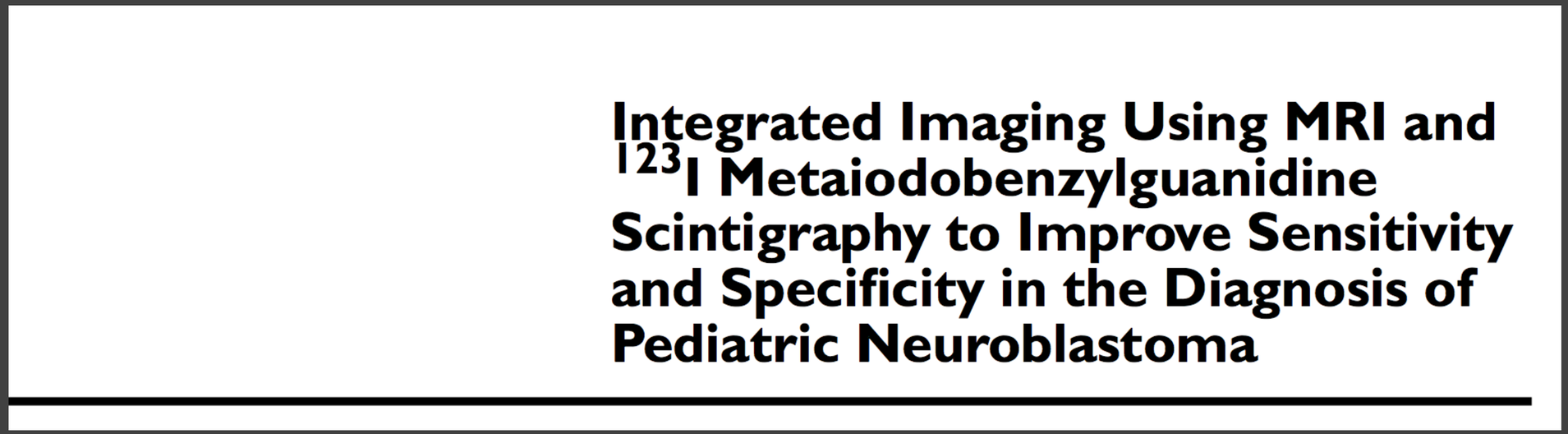
Skull base, calvaria, orbits (25%)

Bone marrow metastases



Virchow node

WB MRI - bone marrow mets?



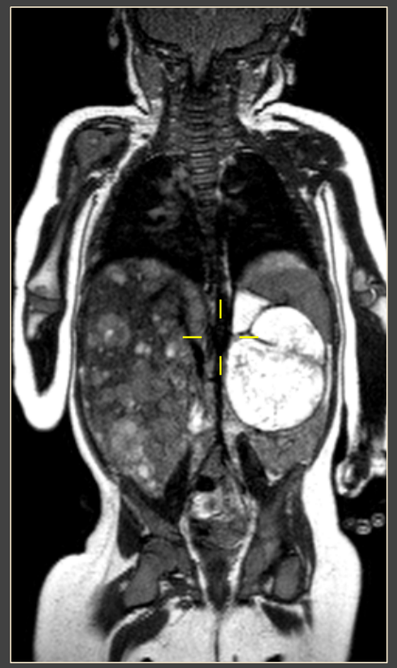
Pfluger et al. AJR 2003

C/ Intergrated imaging showed highest sensitivity and specificity

WB MRI - bone marrow mets?

- Highly sensitive for staging (+DWI)
- Role in MIBG negative tumors?
- Criteria for response of bone marrow lesions?
  - Signal changes persist longer than presence of malignant cells (~lymphoma)
  - Role of ADC value? – how reliable can we measure? Cut off value?

Normal red bone marrow vs diffuse mets?



6 months old with MS

4 months later

Take home messages

Diagnostic features: growth pattern, calcifications & diffusion restriction

Staging: IDRFs

Important time points: pre-operative assessment and pre-RT assessment

Role of MRI in bone marrow metastases?

Thank you for your attention