Neuroradiological Imaging Techniques in Pediatric Neurology

Rajan Patel, MD
Director, Pediatric Neuroimaging
Assistant Professor, Division of Neuroradiology
DISCLOSURE

• No financial disclosure.
LEARNING OBJECTIVES

• Overview of different imaging modalities in Pediatric Neuroradiology

• To learn the basic differences in CT & MRI of the Brain in Pediatrics as compare to adults.

• Brief discussion of role of imaging in Hypoxic Ischemic Encephalopathy (HIE), Seizure, Metabolic Disorders
IMAGING MODALITIES

• **USG:**
  – Brain: Evaluate for IVH, hydrocephalus for preterm and newborn (<6 months)
  – Spine: Spinal Dysraphism, concern for tethered cord

• **CT:**
  – Brain: Screening exam in acute setting/ER with neurological deficit, Trauma, suspected child abuse

• **MR:**
  – Neurological deficit, Developmental delay, Suspected congenital abnormality, Metabolic Disorders.
Head Ultrasound – Why?

- No radiation
- Can be performed at bed-side
- Non-invasive
- Requires no sedations
- Relatively inexpensive
- No contraindications
Head Ultrasound - Limitations

• Operator dependent

• Age of child
  – Can’t be performed once anterior fontanelle closed.

• Limited evaluation for early parenchymal abnormalities
Head Ultrasound - Indications

• Germinal matrix hemorrhage
  - Preterm infants
• Periventricular Leukomalacia (PVL)
• Macrocephaly
• Follow up hydrocephalus
• Concern for presence of vascular anomalies – Vein of Galen Malformation
Watershed Zones: Pre-term Vs. Full term

- Penetrating arteries that arise from the surface of the brain and extend toward lateral ventricles.
- Lack of collaterals.

<table>
<thead>
<tr>
<th>Pre-Term</th>
<th>Full Term</th>
</tr>
</thead>
</table>

- Vessels start to grow from the lateral ventricles outward toward the surface.
- Moves the watershed area more peripherally.
- Difficulty with autoregulation in response to hypoxia or ischemia.

Patterns of brain injury in HIE

• **Mild to moderate HIE:**
  – Blood flow is shunted from the watershed areas to the more hypervascular areas of the brain including the basal ganglia, thalamus, brainstem.
  – Classic periventricular WM findings in preterm and deep WM in full term.

• **Severe HIE:**
  – Deep brain structures such as basal ganglia involvement.
HIE in the Pre-term

• Neuroimaging findings of the HIE depend on the severity of the insult:

  – Mild to moderate HIE:
    • Germinal matrix hemorrhage
    • Intraventricular hemorrhage (IVH)
    • Periventricular Leukomalacia (PVL)

  – Severe HIE:
    • Injuries involving the deep gray matter structures (basal ganglia) or brainstem, which are similar to findings of HII in term infants

Huang BY, Castillo M. Hypoxic-Ischemic Brain Injury: Imaging Findings from Birth to Adulthood. Radiographics 2008
Germinal Matrix Hemorrhage and IVH

Grade I: Hemorrhage confined to caudothalamic groove
Grade II: Grade I + extension into the lateral ventricles without hydrocephalus.
Germinal Matrix Hemorrhage and IVH

Sagittal

Coronal

Grade III: Grade II with hydrocephalus
Germinal Matrix Hemorrhage and IVH

Grade IV: Grade III + extension into the adjacent parenchyma/hemorrhagic venous infarcts
• 5% of infants born before 32 weeks and 15-20% of infants born before 28 weeks will develop cerebral palsy\(^{(1)}\).

• PVL is an important prognostic sign as more than 50% of infants with exhibit some form of cerebral palsy or cognitive/behavioral deficit\(^{(2)}\).

• 2 Types of PVL:
  • Focal PVL will be more associated with cerebral palsy
  • Diffuse PVL will be more associated with cognitive/behavioral deficit

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PERIVENTRICULAR LEUKOMALACIA (PVL)
Spine Ultrasound

- Best in newborn/early infancy
- Limited value after 6 months due to ossification of bony elements
Spine Ultrasound - Indications

• Evaluation of lumbosacral stigmata known to be associated with spinal dysraphism
  – Midline or paramedian soft tissue masses
  – Hair tufts
  – Hemangiomas
  – Paramedian, Deep sacral dimples

• Evaluation of suspected defects such as Cord Tethering, Diastematomyelia, Syrinx
Spine Ultrasound - Indications
Spine Ultrasound - Indications
Radiation Risk

http://apps.who.int/iris/bitstream/10665/205033/1/9789241510349_eng.pdf?ua=1
http://www.acr.org/Quality-Safety/Appropriateness-Criteria
Radiation Risk

**Worldwide average radiation exposure (mSv)**
Total: 3 mSv

- Artificial sources other than medical (0.3%) → 0.01
- Radon (41.7%) → 1.26
- Natural sources other than radon (38.3%) → 1.14
- Medical exposure (19.7%) → 0.6

**US average radiation exposure (mSv)**
Total: 6.11 mSv

- Artificial sources other than medical (0.3%) → 0.15
- Radon (33%) → 1.98
- Natural sources other than radon (16%) → 0.98
- Medical exposure (~50%) → 3.0

*Source: Adapted, with permission, from UNSCEAR (2010)*

*Source: Adapted, with permission, from NCRP (2009)*
Radiation Risk

Source: Adapted, with permission, from NCRP (2009)
## Radiation Risk

<table>
<thead>
<tr>
<th>X-ray examination</th>
<th>Relative frequency (%)</th>
<th>Collective dose (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chest examinations (PA, lateral, others)</td>
<td>40</td>
<td>13.3</td>
</tr>
<tr>
<td>Limb and joint</td>
<td>8.4</td>
<td>&lt; 1</td>
</tr>
<tr>
<td>Skull</td>
<td>3.2</td>
<td>4.2</td>
</tr>
<tr>
<td>Abdomen, pelvis, hip</td>
<td>5.2</td>
<td>4.5</td>
</tr>
<tr>
<td>Spine</td>
<td>7.4</td>
<td>4.2</td>
</tr>
<tr>
<td>Fluoroscopic studies of the gastrointestinal tract</td>
<td>4.8</td>
<td>14.5</td>
</tr>
<tr>
<td>Mammography</td>
<td>3.6</td>
<td>&lt; 1</td>
</tr>
<tr>
<td><strong>Computed tomography</strong></td>
<td><strong>6.3</strong></td>
<td><strong>43.2</strong></td>
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<tr>
<td>Angiography and fluoroscopy-guided interventional procedures</td>
<td>&lt; 1</td>
<td>6.1</td>
</tr>
<tr>
<td>Other X-ray medical imaging procedures</td>
<td>3</td>
<td>11</td>
</tr>
<tr>
<td>Dental procedures&lt;sup&gt;c&lt;/sup&gt;</td>
<td>13</td>
<td>&lt; 1</td>
</tr>
</tbody>
</table>
## Radiation Risk

<table>
<thead>
<tr>
<th>Diagnostic procedure</th>
<th>Equivalent number of chest X-rays</th>
<th>Equivalent period of exposure to natural radiation</th>
<th>Typical effective dose (mSv)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Chest X-ray</strong> (single PA film)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adult</td>
<td>1</td>
<td>3 days</td>
<td>0.02&lt;sup&gt;c&lt;/sup&gt;</td>
</tr>
<tr>
<td>5-year-old</td>
<td>1</td>
<td>3 days</td>
<td>0.02&lt;sup&gt;c&lt;/sup&gt;</td>
</tr>
<tr>
<td><strong>CT head</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Adult</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Newborn</td>
<td>200</td>
<td>2.5 years</td>
<td>6</td>
</tr>
<tr>
<td>1-year-old</td>
<td>185</td>
<td>1.5 years</td>
<td>3.7</td>
</tr>
<tr>
<td>5-year-old</td>
<td>100</td>
<td>10 months</td>
<td>2&lt;sup&gt;d&lt;/sup&gt;</td>
</tr>
<tr>
<td>10-year-old</td>
<td>110</td>
<td>11 months</td>
<td>2.2</td>
</tr>
<tr>
<td>Paediatric head CT angiography&lt;sup&gt;f&lt;/sup&gt;</td>
<td>250</td>
<td>2 years</td>
<td>5</td>
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</table>
Decreased Dose

- kVp>mA>collimation.
  - Increase mA with age.
    - 0 to 2 years 135.
    - 2-3 years 180.
    - 3-6 years 210.
    - > 6 years 240

- Low dose CT for shunt evaluation, PNS, Temporal bone

- Use other alternatives if possible such as MRI for shunt evaluation.
Unique about Pediatrics CT HEAD

• White matter is difficult to evaluate in the neonate
  – Better evaluated with US and MR

• Marked white matter hypodensity until age of 2 months

• Basal ganglia indistinct until 4 months

• Cortex should be well seen no matter what age

• Hyperdense dural venous sinuses in neonates from increased hematocrits

• Prominence of the subarachnoid spaces
  – Normal or macrocephalic
3 week old with Seizure
7 week old with Fever
3 year old with Seizure
3 week old with Seizure
3 year old with Seizure, Weakness

Which side is abnormal?
3 week old with Altered consciousness
3 week old with Fever, Seizure
14 month old with macrocephaly

- Normal or Abnormal
- Which compartment fluid is located?
6 month old with macrocephaly

- Normal or Abnormal
- Which compartment fluid is located?
Case 1 - Normal or Abnormal
Which compartment fluid is located?

Case 2
Anterior Extra-axial Fluid

• Subarachnoid versus Subdural
  • Subarachnoid
    – Symmetric
    – Vessels (bridging veins) coarse through
    – No mass effect
  • Subdural
    – Asymmetric
    – Vessels displaced towards cortex
    – Underlying mass effect.
4 months, Girl with 1\textsuperscript{st} time seizure & 5 mins of passing out, CPR by Dad

- **Social History:**
  - Wanted child – only child
  - Socially well adjusted family

- **No History of trauma**

- **Unclear diagnosis!**
  - Epilepsy? – Strong family history
  - Cardiac problem?
  - Metabolic etiology?
4 months, Girl with 1\textsuperscript{st} time seizure & 5 mins of passing out, CPR by Dad

- Clinical examination:
  - Bruise on the both sides of chest
  - H/o CPR by Dad at home
Disease/accident vs. Abuse - a question of balance?

- Socially well adjusted family
- Seizure without trauma
- No report of trauma
- CT report: Normal
- Disease/Accident
- Abuse
4 months, Girl with 1<sup>st</sup> time seizure & 5 mins of passing out, CPR by Dad
Disease/accident vs. Abuse
- a question of balance?

Seizure without trauma
Socially well adjusted family

Disease/Accident

CT report: Clear!
- Bilat. SDH suggesting violent, recurring trauma
Positive Retinal Hemorrhage

Abuse
SDH: Time course Vs. Change of attenuation (HU)

Adapted from K. Ericson
Child Abuse

- Retinal haemorrhages
- Acute on chronic SDH
- Cerebral edema
- Speaks in very strong favour

Shaken Baby Syndrome (> 95 %)

Bridging Veins
SHAKEN BABY SYNDROME

- Repeated accelerations – decelerations
- Brain comes in motion relative to the skull and meninges.
- Torn bridging veins
- Subdural haematoma
Pediatrics MRI HEAD
White Matter Myelination

- **T1** – WM *increases* relative to GM
- **T2** – WM *decreases* relative to GM
White Matter Myelination

Caudal to Cranial

Central to Peripheral

Posterior to Anterior
### Simplified Pattern Approach

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<th>T2</th>
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Up to 6 months: Use T1-W; Use T2-W > 6 months
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Up to 6 months: Use T1-W; Use T2-W > 6 months
T1 Weighted Sequence

- MCP, DCW
- Splenium
- Genu

- 3 months
- 4 months
- 6 months
- 8-11 months

T2 Weighted Sequence

- Splenium
- Genu
- ALIC

- 6 months
- 8 months
- 11 months
- 14-18 months
Role of imaging in Epilepsy

• To identify possible epileptic focus
  – MRI, MEG, SPECT/PET

• To confirm lateralization
  – fMRI, WADA testing
MRI: EPILEPSY PROTOCOL

- Imaging MUST BE acquired on a 3T scanner.
- High resolution T2-weighted coronal oblique images
- 3D T1-weighted MPRAGE volumetric images
- Coronal FLAIR images
WHY 3T?

1.5 T, Coronal T2 Oblique Images
3 T, Coronal T2 Oblique Images

- Abnormally thickened right insular cortex
- Abnormally thickened superior temporal gyrus
  - Right temporal subependymal heterotopia.
<table>
<thead>
<tr>
<th>CAUSE</th>
<th>0-2</th>
<th>2-20</th>
<th>21-40</th>
<th>41-60</th>
<th>&gt;61</th>
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<tbody>
<tr>
<td>Cerebral Hypoxia</td>
<td>X</td>
<td></td>
<td></td>
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<tr>
<td>Inborn error of Metabolism</td>
<td>X</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Congenital Malformation</td>
<td>X</td>
<td>X</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Infection</td>
<td>X</td>
<td>X</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Phakomatosis</td>
<td>X</td>
<td>X</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Primary seizures</td>
<td></td>
<td>X</td>
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</tr>
<tr>
<td>MTS</td>
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<td>X</td>
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<tr>
<td>Vascular Malformation</td>
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<tr>
<td>Post-traumatic Epilepsy</td>
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<tr>
<td>Tumor</td>
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<tr>
<td>Stroke</td>
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MALFORMATION OF CORTICAL DEVELOPMENT

• **Malformation due to abnormal neuronal and glial proliferation**
  - Focal cortical dysplasia
  - Hemimegalencephaly
  - Neoplasm (DNET, Ganglioglioma)

• **Malformation due to abnormal neuronal migration**
  - Lissencephaly
  - Heterotopias

• **Malformation due to abnormal late migration and organization**
  - Polymicrogyria
  - Schizencephaly
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• Key MRI Features of FCD:
  – Abnormal gyral pattern
FCD: KEY MR IMAGING FINDINGS

• Key MRI Features of FCD:
  – Abnormal gyral pattern
  – Increased cortical thickness
  – Increased cortical signal
FCD: KEY MR IMAGING FINDINGS

• Key MRI Features of FCD:
  – Abnormal gyral pattern
  – Increased cortical thickness
  – Increased cortical signal
  – Blurring of gray matter / white matter junction
  – Increased white matter signal
FCD: KEY MR IMAGING FINDINGS

- Abnormal gyral pattern
- Increased cortical thickness
- Increased cortical signal
- Blurring of gray matter / white matter junction
- Increased white matter signal
- ‘Transmantle’ signal changes
Mesial Temporal Sclerosis (MTS)

- Left hippocampus \textbf{T2 hyperintensity}
- Left hippocampal \textbf{atrophy}
- Compensatory \textbf{enlargement of the adjacent temporal horn}

\textbf{Coronal T2}
Pediatric Metabolic Disorders

- Genetic Tests
- Clinical Data
- Lab Tests
- Imaging
Pediatric Metabolic Disorders

- Genetic Tests
- Clinical Data
- Lab Tests
- Imaging
MRI based approach to Pediatric Metabolic Ds.

1) Disorders of Hypomyelination
2) Disorders affecting subcortical WM
3) Disorders affecting periventricular + deep WM
4) Disorders affecting combined WM and GM
5) Disorders affecting GM with some WM
Disorders of Hypomyelination

- Pelizaeus–Merzbacher disease (PMD)
- Fucosidosis
- Sialic acid storage disorder
- Hypomyelination with atrophy of the basal ganglia and cerebellum
- Hypomyelination, hypodontia, hypogonadotropic hypogonadism
- Hypomyelination and congenital cataract
- Hypomyelination with monocarboxylate transporter-8 deficiency
- Folate receptor defect
- Tremor-ataxia with central hypomyelination
Markedly delayed myelination of the cerebral white matter on T2.
*Disorders affecting Predominantly Subcortical WM*

- Megalencephalic Leukoencephalopathy with Cysts (MLC) / Van der Knaap Disease
- Vanishing White Matter Disease
- Galactosemia
- Aicardi-Goutieres Syndrome
Vanishing White Matter Ds.
(Childhood ataxia with central hypomyelination)
Megalencephalic Leukoencephalopathy with Cysts (MLC) / Van der Knaap Disease
*Disorders affecting Predominantly Periventricular + Deep WM*

- X-linked ALD
- Metachromatic Leukodystrophy
- Mucopolysaccharidoses (MPS)
- Lowe Syndrome
- Cockayne Syndrome
- X-linked Charcot-Marie-Tooth Syndrome
- Leukoencephalopathy with Brainstem and Spinal cord involvement (LBSL)
X-Linked Adrenoleukodystrophy (ALD)
Metachromatic Leukodystrophy
Disorders affecting Combined White and Gray Matter

- Alexander Disease
- Canavan Disease
- Krabbe Disease
- Maple Syrup Urine Disease (MSUD)
- GM1/GM2 Gangliosidosis
- Organic Acidopathies
  - e.g. Propionic acidemia, Glutaric aciduria type I, Methylmalonic acidemia, L-2-hydroxyglutaric aciduria
Alexander Disease
Canavan Disease

13 months old, increased head circumference
Canavan Disease

Elevated NAA : Creatine ratio 2.72
*Disorders affecting Predominantly Gray Matter with some WM*

- Mitochondrial Disorders
  - Leigh Syndrome
  - Kearns-Sayre Syndrome
  - Mitochondrial Encephalopathy, Lactic Acidosis, Stroke like symptoms (MELAS)
- Urea Cycle Disorders
Leigh Syndrome
SUMMARY

• Use helpful resources if any uncertainty regarding which imaging study to order in pediatric patients.
  – ACR appropriate criteria, Call Radiologists

• Significant difference in appearance of CT Head (up to 6 months) & MRI of head (up to 2 year) as compared to adults.

• Learn from your colleagues.
  i.e. multidisciplinary conferences
Thank you for your attention

Rajan.P.Patel@uth.tmc.edu