Neuroblastomas
Image Defined Risk Factors

H.J. Brisse, Paris, France
INRG Staging System

Why ?

- Neuroblastoma: a very heterogeneous disease
- Numerous prognostic factors and treatment options
- Risk-based treatment approach

➢ Need for a pre-treatment staging system
## Local disease staging

### INSS
**depends on surgery**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Quality of resection</th>
<th>Non-adherent ipsilateral LN</th>
<th>Contralateral LN</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>R0 R1</td>
<td>N0</td>
<td>N0</td>
</tr>
<tr>
<td>2A</td>
<td>R2</td>
<td>N0</td>
<td>N0</td>
</tr>
<tr>
<td>2B</td>
<td>R0 R1 R2</td>
<td>N+</td>
<td>N0</td>
</tr>
<tr>
<td>3</td>
<td>« Unresectable » Cross midline</td>
<td>N0 N+</td>
<td>N0 N+</td>
</tr>
</tbody>
</table>

### INRG SS
**Before any treatment**

<table>
<thead>
<tr>
<th>Stage</th>
<th>Image Defined Risk Factors</th>
</tr>
</thead>
<tbody>
<tr>
<td>L1</td>
<td>IDRF = 0</td>
</tr>
<tr>
<td>L2</td>
<td>IDRF ≥ 1</td>
</tr>
</tbody>
</table>

Monclair T, et al. JCO 2009

Image Defined Risk Factors

IDRFs are

- Surgical risk factors
- Identified by imaging
- at diagnosis, before any treatment
- based on radiological criteria
- defined by international consensus
- allowing consistent and uniform reporting

IDRFs are NOT

- Contraindication for surgery
- Definitive criteria

References:

Ceccheto G et al. JCO 2005 (LNESG1)

Monclair T, et al. JCO 2009
Brisse HJ et al. Radiology 2011
Pohl A et al. Klinishe Padiatrie 2016
INRG Imaging Committee

- **Pediatric radiologists**
  - Hervé Brisse\textsuperscript{3,5} (Chair)
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  - Claudio Granata\textsuperscript{5}
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  - Barabara Krug\textsuperscript{2}
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  - Val Lewington\textsuperscript{5}

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  - Andrew Pearson\textsuperscript{3}

- **Statistician**
  - Wendy London\textsuperscript{3}

from COG\textsuperscript{1}, GPOH\textsuperscript{2}, INRG\textsuperscript{3}, JNBSG\textsuperscript{4}, SIOPEN\textsuperscript{5}
### IDRF Check-list

**Checklist to ensure correct INRGSS Staging of Patients with Neuroblastic Tumors**

Section A and B to be completed by the radiologist(s). Section C (if used) to be completed by the multidisciplinary treatment team. Every row of the form should be completed with either a “Yes”, “No” or “N.A.” (Not Assessed or Not Assessable). INRGSS – Image Defined Risk Factors

#### Section A

**EXTENT OF PRIMARY TUMOR - IDRF**

<table>
<thead>
<tr>
<th></th>
<th>Yes</th>
<th>No</th>
<th>N.A.</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>Ipsilateral tumor extension within two body compartments</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>A.1 Neck-chest</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>A.2 Chest-abdomen</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>A.3 Abdomen-pelvis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>B</td>
<td>Neck</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>B.1 Tumor encasing carotid and/or vertebral artery and/or internal jugular vein</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>B.2 Tumor extending to base of skull</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>B.3 Tumor compressing the trachea</td>
<td></td>
<td></td>
</tr>
<tr>
<td>C</td>
<td>Cervico-thoracic junction</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>C.1 Tumor encasing brachial plexus roots</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>C.2 Tumor encasing subclavian vessels and/or vertebral and/or carotid artery</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>C.3 Tumor compressing the trachea</td>
<td></td>
<td></td>
</tr>
<tr>
<td>D</td>
<td>Thorax</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>D.1 Tumor encasing the aorta and/or major branches</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>D.2 Tumor compressing the trachea and/or principal bronchi</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>D.3 Lower mediasinal tumor, mitigating the costo-vertebral junction between T9 and T12</td>
<td></td>
<td></td>
</tr>
<tr>
<td>E</td>
<td>Thoraco-abdominal</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>E.1 Tumor encasing the aorta and/or vena cava</td>
<td></td>
<td></td>
</tr>
<tr>
<td>F</td>
<td>Abdomen-pelvis</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F.1 Tumor infiltrating the porta hepatica (liver hilum) and/or hepatoduodenal ligament</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F.2 Tumor encasing branches of the superior mesenteric artery at the mesenteric root</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F.3 Tumor encasing the origin of the celiac axes and/or of the superior mesenteric artery</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F.4 Tumor invading one or both renal pedicles</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F.5 Tumor encasing the aorta and/or vena cava</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F.6 Tumor encasing the iliac vessels</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>F.7 Tumor crossing the aortic notch</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**IDRF status of the primary tumor at diagnosis:**

**IDRF Negative** – All rows were checked “No” or “N.A.”

**IDRF Positive** – One or more rows were checked “Yes.”

### Section B

**Conditions to be recorded but not considered IDRFs**

<table>
<thead>
<tr>
<th></th>
<th>Yes</th>
<th>No</th>
<th>N.A.</th>
</tr>
</thead>
<tbody>
<tr>
<td>J</td>
<td>Multifocal primary tumors</td>
<td></td>
<td></td>
</tr>
<tr>
<td>J.2</td>
<td>Right-sided pleural effusion</td>
<td></td>
<td></td>
</tr>
<tr>
<td>J.3</td>
<td>Left-sided pleural effusion</td>
<td></td>
<td></td>
</tr>
<tr>
<td>J.4</td>
<td>Ascites</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Date**

**Radiologist 1**

**Radiologist 2 (Optional)**
Definitions of terms

« Contact »

- **Artery**
  - Contact (< 50%)

- **Vein**
  - flattened (visible lumen)

- **NB**
  - No IDRF

« Encasement »

- **Artery**
  - >50%

- **Vein**
  - flattened (no visible lumen)

- **NB**
  - Total encasement

- **Partial encasement**
  - (contact ≥ 50%)

- **IDRF**
  - No IDRF

IDRF

Institut Curie - HJ Brisse – Oncology & Nuclear Medicine – Neuroblastoma

IPR - Chicago – May 2016
Chest

IDRF=0

IDRF+

IDRF+
Pelvis

IDRF=0

IDRF +

IDRF +
Neck

IDRF = 0

IDRF +

IDRF +

IDRF +
Take Home Message

IDRF helps

Radiologists: relevant reporting
Pediatricians: local staging / trial treatment arms
Surgeons: risk of complication or incomplete removal
Statisticans: standardization / trials comparison

Major criteria

- Multicompartmental tumor
- Encasement of large vessels
- Contiguous organs infiltration
- Significant intraspinal extension
- Airway compression