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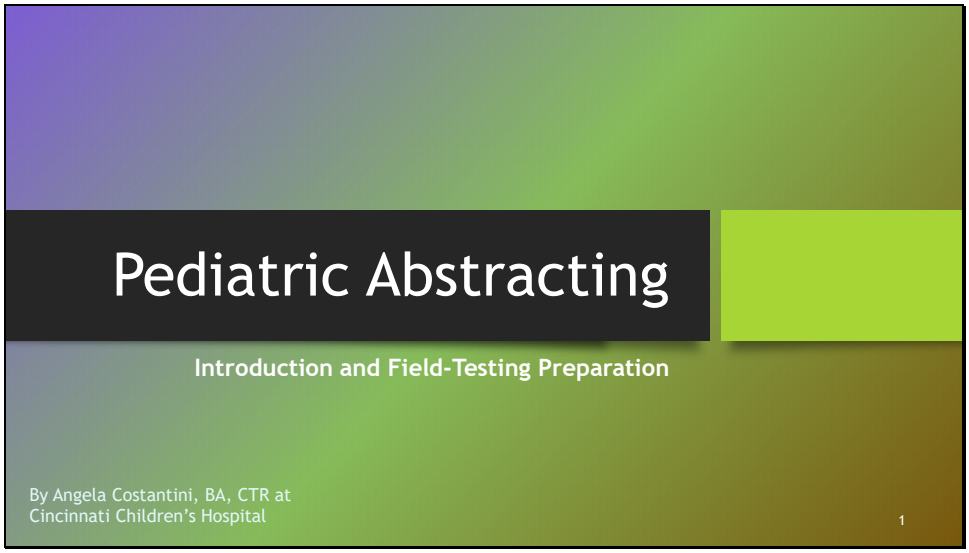
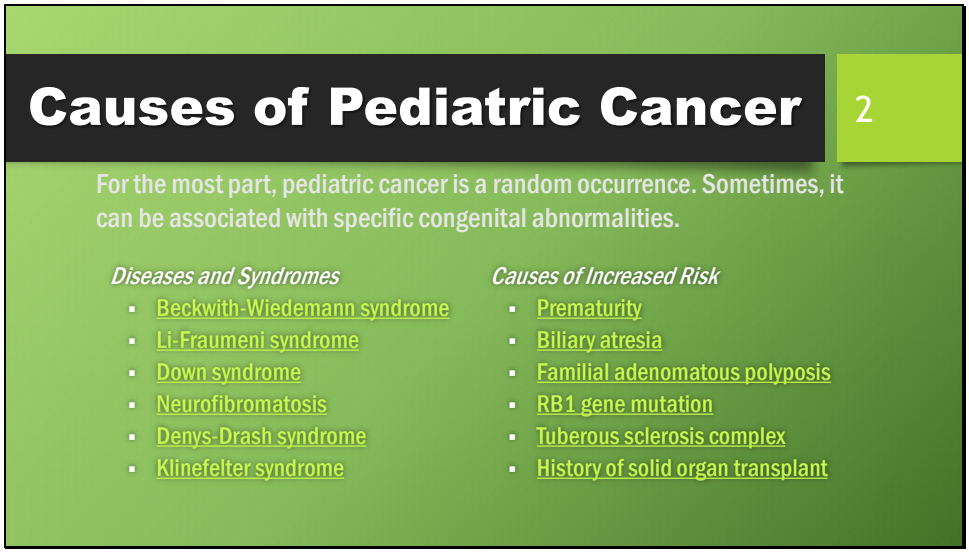
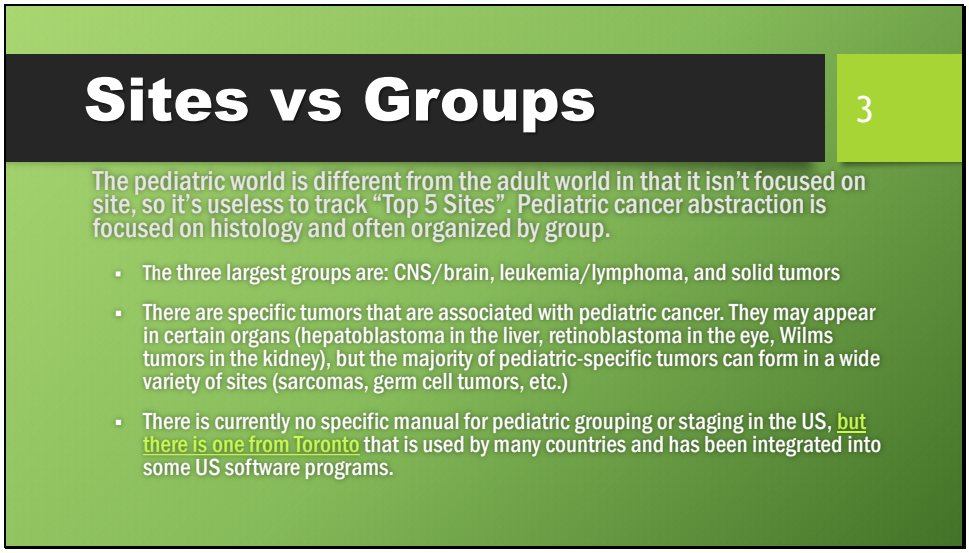

Pediatric Abstracting
Introduction and Field-Testing Preparation
By Angela Costantini, BA, CTR at
Cincinnati Children's Hospital
1

Slide 2


Causes of Pediatric Cancer
2
For the most part, pediatric cancer is a random occurrence. Sometimes, it can be associated with specific congenital abnormalities.
Diseases and Syndromes
Beckwith-Wiedemann syndrome
Li-Fraumeni syndrome
Down syndrome
Neurofibromatosis
Denys-Drash syndrome
Klinefelter syndrome
Causes of Increased Risk
Prematurity
Biliary atresia
Familial adenomatous polyposis
RB1 gene mutation
Tuberous sclerosis complex
History of solid organ transplant

Slide 3


Sites vs Groups
3
The pediatric world is different from the adult world in that it isn't focused on site, so it's useless to track "Top 5 Sites". Pediatric cancer abstraction is focused on histology and often organized by group.
The three largest groups are: CNS/brain, leukemia/lymphoma, and solid tumors
There are specific tumors that are associated with pediatric cancer. They may appear in certain organs (hepatoblastoma in the liver, retinoblastoma in the eye, Wilms tumors in the kidney), but the majority of pediatric-specific tumors can form in a wide variety of sites (sarcomas, germ cell tumors, etc.)
There is currently no specific manual for pediatric grouping or staging in the US, but there is one from Toronto that is used by many countries and has been integrated into some US software programs.

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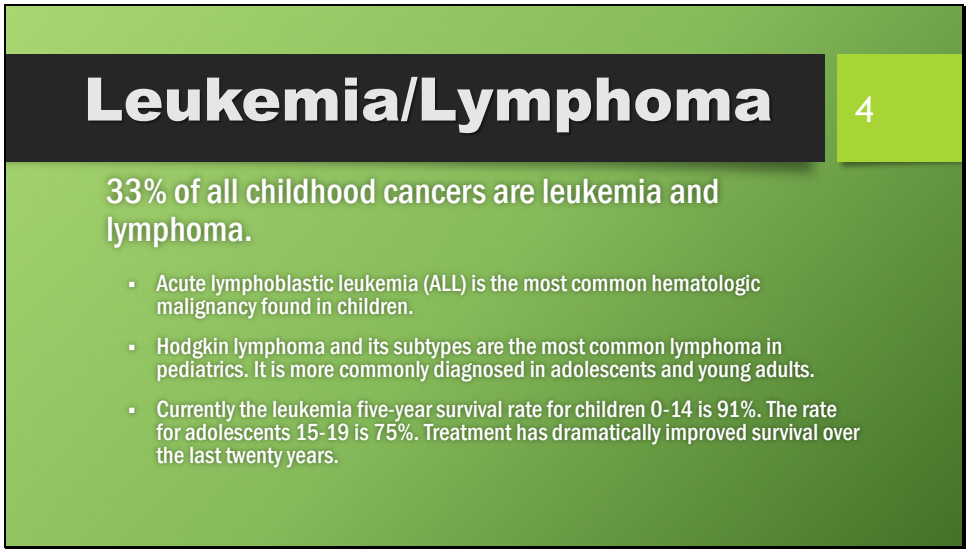
Leukemia/Lymphoma
4
33% of all childhood cancers are leukemia and lymphoma.
Acute lymphoblastic leukemia (ALL) is the most common hematologic malignancy found in children.
Hodgkin lymphoma and its subtypes are the most common lymphoma in pediatrics. It is more commonly diagnosed in adolescents and young adults.
Currently the leukemia five-year survival rate for children 0-14 is 91%. The rate for adolescents 15-19 is 75%. Treatment has dramatically improved survival over the last twenty years.

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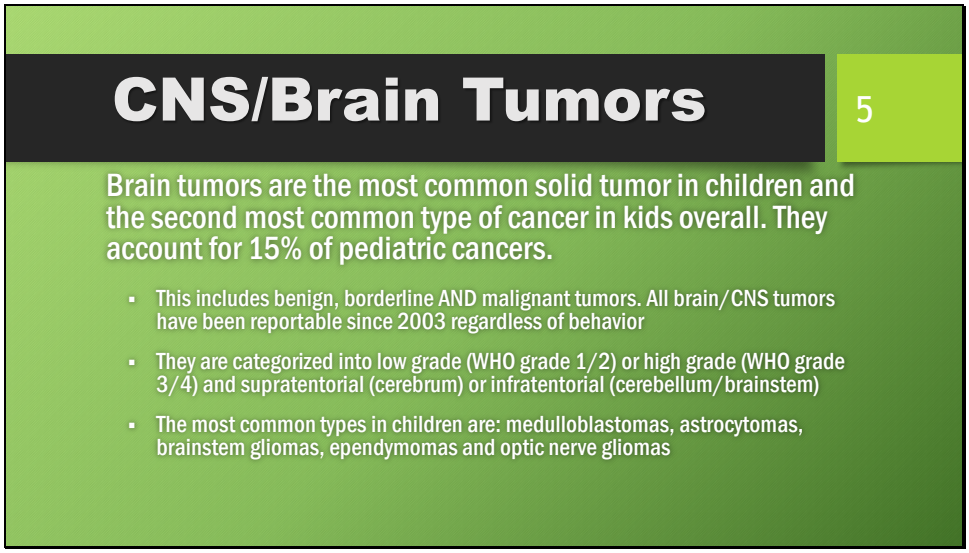
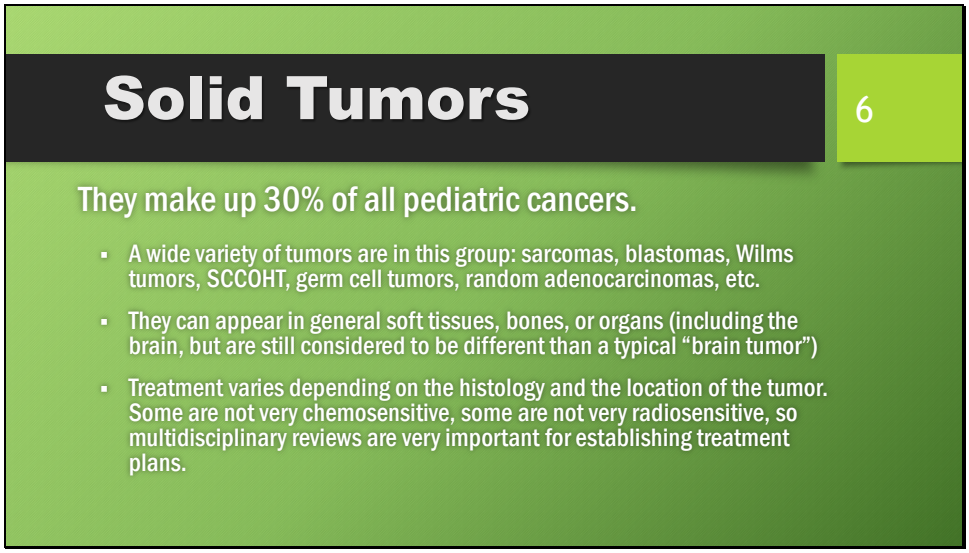
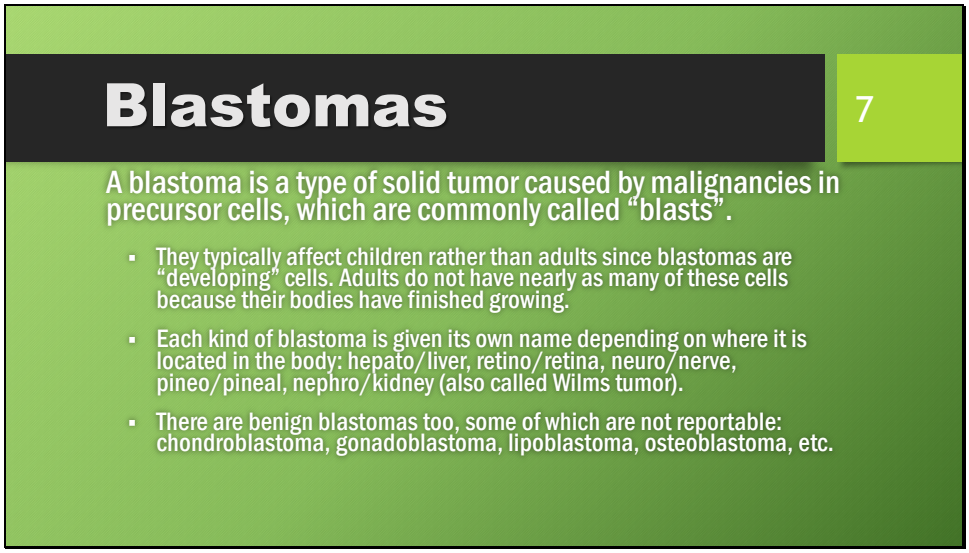

CNS/Brain Tumors
5
Brain tumors are the most common solid tumor in children and the second most common type of cancer in kids overall. They account for 15% of pediatric cancers.
▪ This includes benign, borderline AND malignant tumors. All brain/CNS tumors have been reportable since 2003 regardless of behavior
▪ They are categorized into low grade (WHO grade 1/2) or high grade (WHO grade 3/4) and supratentorial (cerebrum) or infratentorial (cerebellum/brainstem)
▪ The most common types in children are: medulloblastomas, astrocytomas, brainstem gliomas, ependymomas and optic nerve gliomas

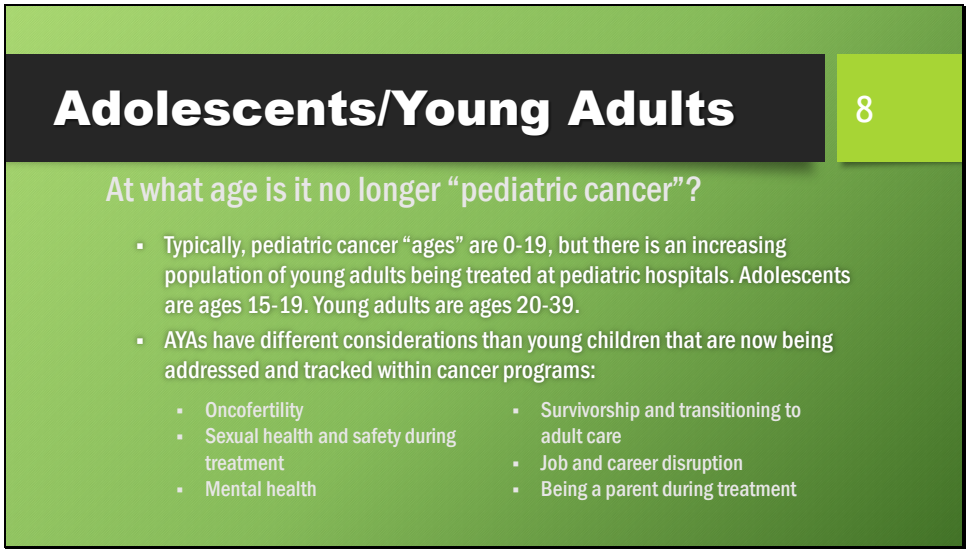
Slide 6


Solid Tumors
6
They make up 30% of all pediatric cancers.
A wide variety of tumors are in this group: sarcomas, blastomas, Wilms tumors, SCCOHT, germ cell tumors, random adenocarcinomas, etc.
They can appear in general soft tissues, bones, or organs (including the brain, but are still considered to be different than a typical “brain tumor”)
Treatment varies depending on the histology and the location of the tumor. Some are not very chemosensitive, some are not very radiosensitive, so multidisciplinary reviews are very important for establishing treatment plans.

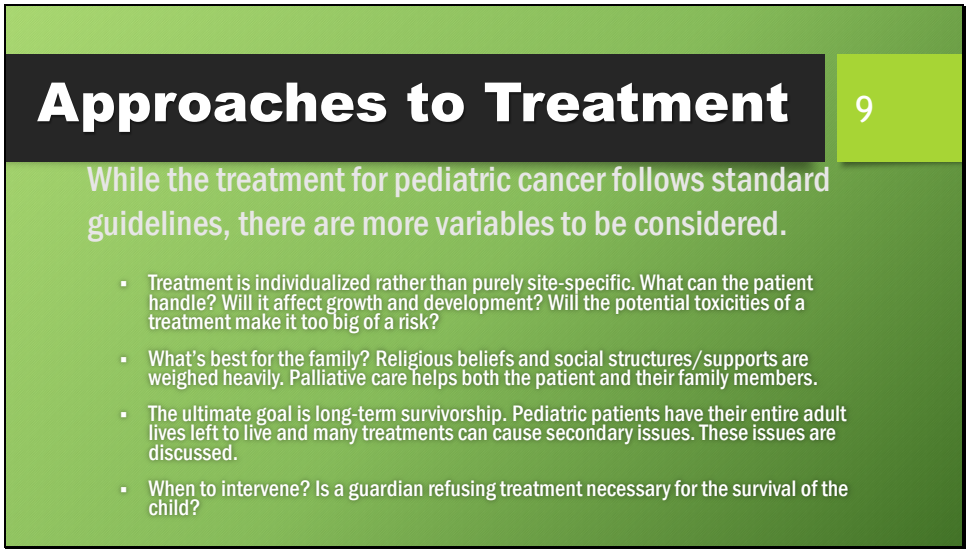
Slide 7


Blastomas
7
A blastoma is a type of solid tumor caused by malignancies in precursor cells, which are commonly called "blasts".
They typically affect children rather than adults since blastomas are "developing" cells. Adults do not have nearly as many of these cells because their bodies have finished growing.
Each kind of blastoma is given its own name depending on where it is located in the body: hepato/liver, retino/retina, neuro/nerve, pineo/pineal, nephro/kidney (also called Wilms tumor).
There are benign blastomas too, some of which are not reportable: chondroblastoma, gonadoblastoma, lipoblastoma, osteoblastoma, etc.

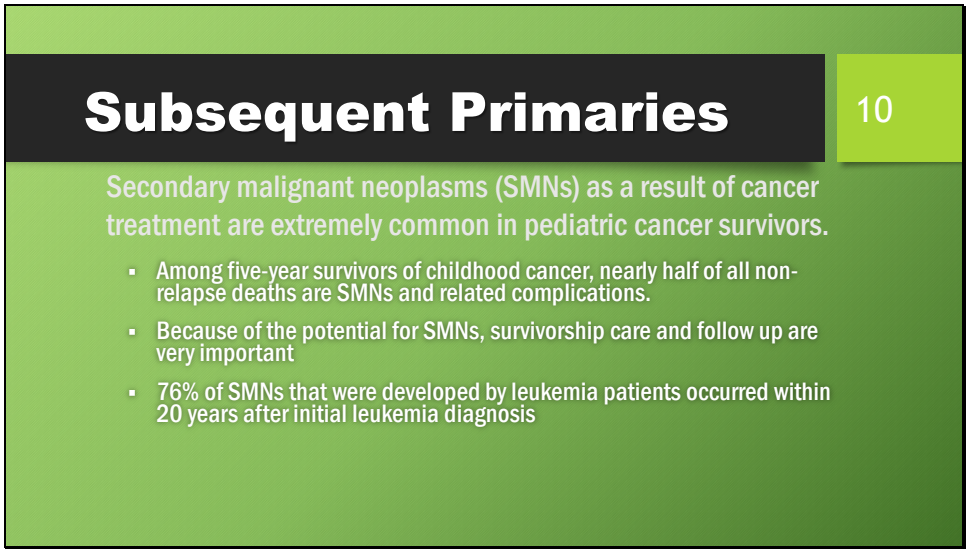
Slide 8


Adolescents/Young Adults
8
At what age is it no longer "pediatric cancer"?
Typically, pediatric cancer "ages" are 0-19, but there is an increasing population of young adults being treated at pediatric hospitals. Adolescents are ages 15-19. Young adults are ages 20-39.
AYAs have different considerations than young children that are now being addressed and tracked within cancer programs:
Oncofertility
Sexual health and safety during treatment
Mental health
Survivorship and transitioning to adult care
Job and career disruption
Being a parent during treatment

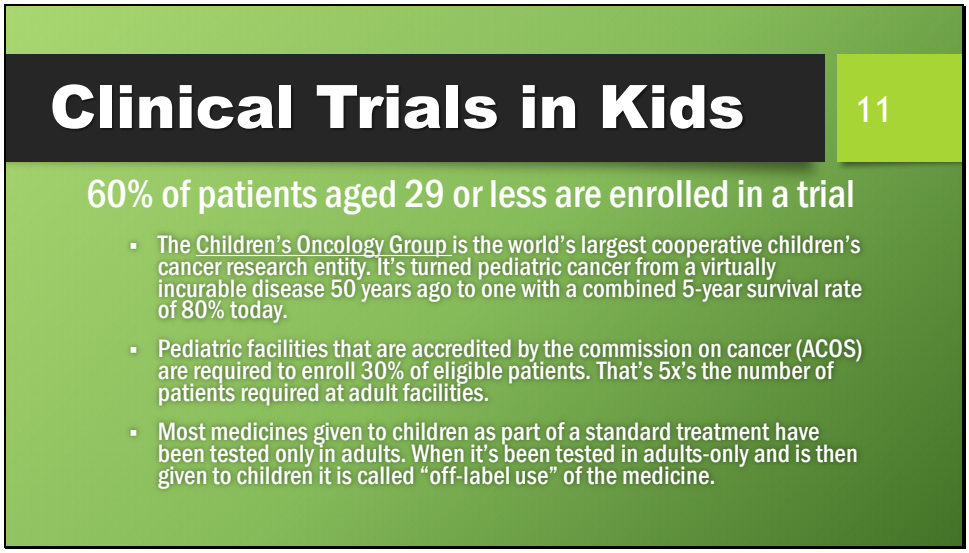
Slide 9


Approaches to Treatment
9
While the treatment for pediatric cancer follows standard guidelines, there are more variables to be considered.
Treatment is individualized rather than purely site-specific. What can the patient handle? Will it affect growth and development? Will the potential toxicities of a treatment make it too big of a risk?
What's best for the family? Religious beliefs and social structures/supports are weighed heavily. Palliative care helps both the patient and their family members.
The ultimate goal is long-term survivorship. Pediatric patients have their entire adult lives left to live and many treatments can cause secondary issues. These issues are discussed.
When to intervene? Is a guardian refusing treatment necessary for the survival of the child?

Slide 10


Subsequent Primaries
10
Secondary malignant neoplasms (SMNs) as a result of cancer treatment are extremely common in pediatric cancer survivors.
Among five-year survivors of childhood cancer, nearly half of all non-relapse deaths are SMNs and related complications.
Because of the potential for SMNs, survivorship care and follow up are very important
76% of SMNs that were developed by leukemia patients occurred within 20 years after initial leukemia diagnosis

Slide 11


Clinical Trials in Kids
11
60% of patients aged 29 or less are enrolled in a trial
The Children's Oncology Group is the world's largest cooperative children's cancer research entity. It's turned pediatric cancer from a virtually incurable disease 50 years ago to one with a combined 5-year survival rate of 80% today.
Pediatric facilities that are accredited by the commission on cancer (ACOS) are required to enroll 30% of eligible patients. That's 5x's the number of patients required at adult facilities.
Most medicines given to children as part of a standard treatment have been tested only in adults. When it's been tested in adults-only and is then given to children it is called "off-label use" of the medicine.

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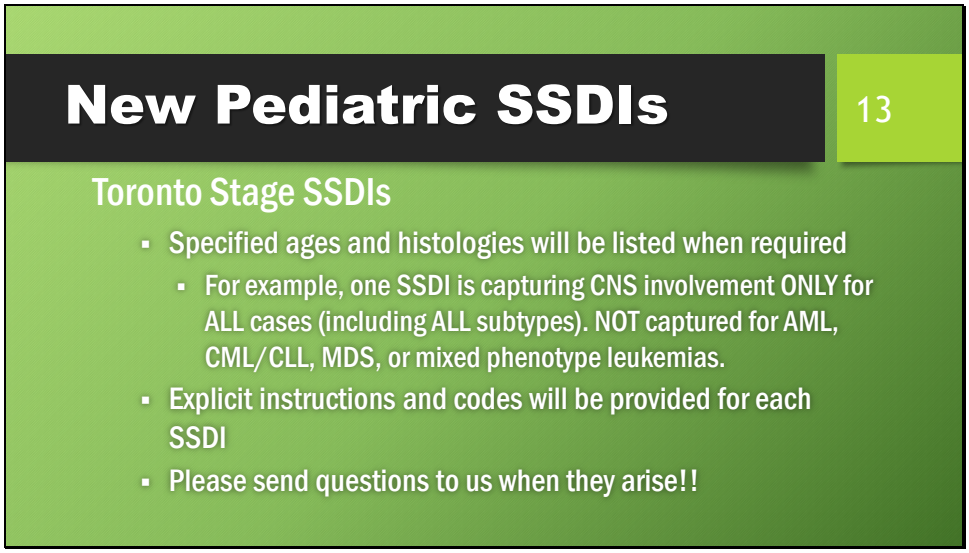
Slide 13


New Pediatric SSDIs
13
Toronto Stage SSDIs
Specified ages and histologies will be listed when required
For example, one SSDI is capturing CNS involvement ONLY for ALL cases (including ALL subtypes). NOT captured for AML, CML/CLL, MDS, or mixed phenotype leukemias.
Explicit instructions and codes will be provided for each SSDI
Please send questions to us when they arise!!

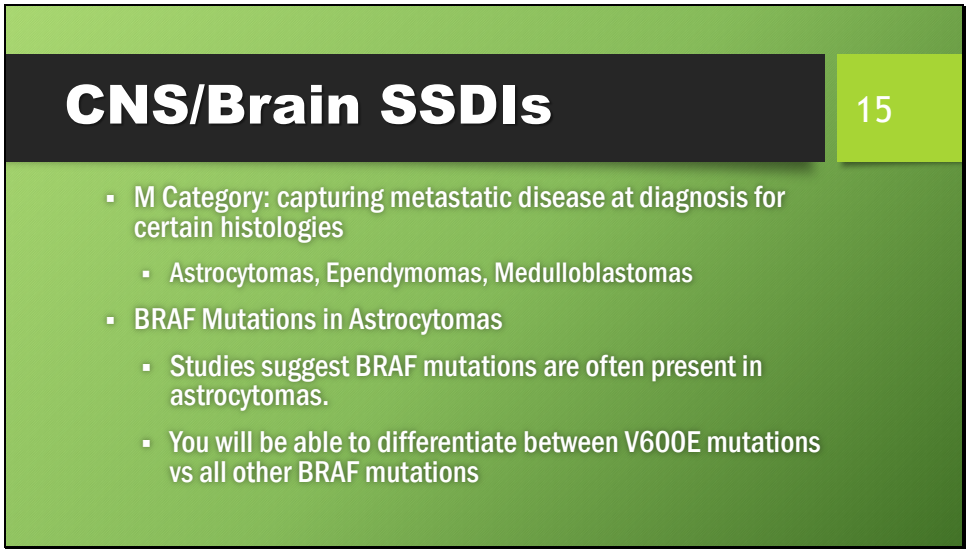
Slide 14

Leuk/Lymph SSDIs

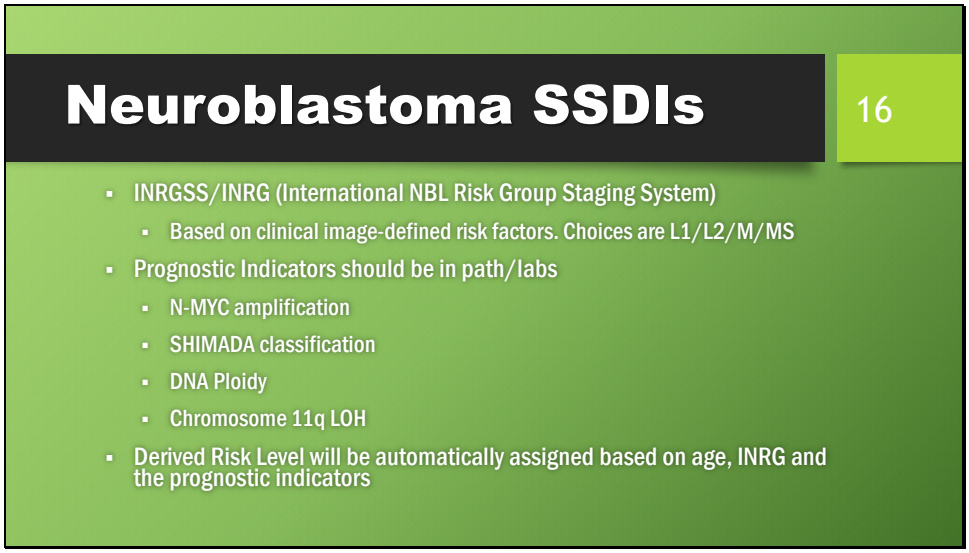
14

- CNS involvement ONLY for ALL cases (including ALL subtypes).
- WBC Count pre-treatment
  - NOT captured for AML, CML/CLL, MDS, or mixed phenotype leukemias for both of the fields above
- Hodgkin Lymphoma cases will get a derived stage based on certain data fields you are already filling out (primary site and B symptoms). You should not have to stage it yourself.
- NHL/Burkitt Lymphoma will get a St. Jude/Murphy Stage at dx. It should be documented by the treating MD, but the stages will be defined in the coding instructions

Slide 15


CNS/Brain SSDIs
15
M Category: capturing metastatic disease at diagnosis for certain histologies
Astrocytomas, Ependymomas, Medulloblastomas
BRAF Mutations in Astrocytomas
Studies suggest BRAF mutations are often present in astrocytomas.
You will be able to differentiate between V600E mutations vs all other BRAF mutations

Slide 16


Neuroblastoma SSDIs
16
INRGSS/INRG (International NBL Risk Group Staging System)
Based on clinical image-defined risk factors. Choices are L1/L2/M/MS
Prognostic Indicators should be in path/labs
N-MYC amplification
SHIMADA classification
DNA Ploidy
Chromosome 11q LOH
Derived Risk Level will be automatically assigned based on age, INRG and the prognostic indicators

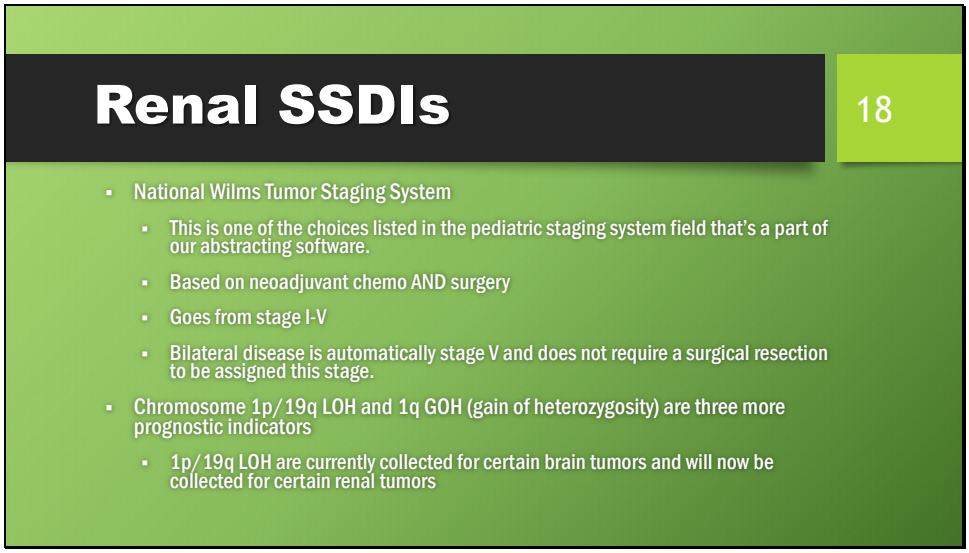
Slide 17

Retinoblastoma SSDIs

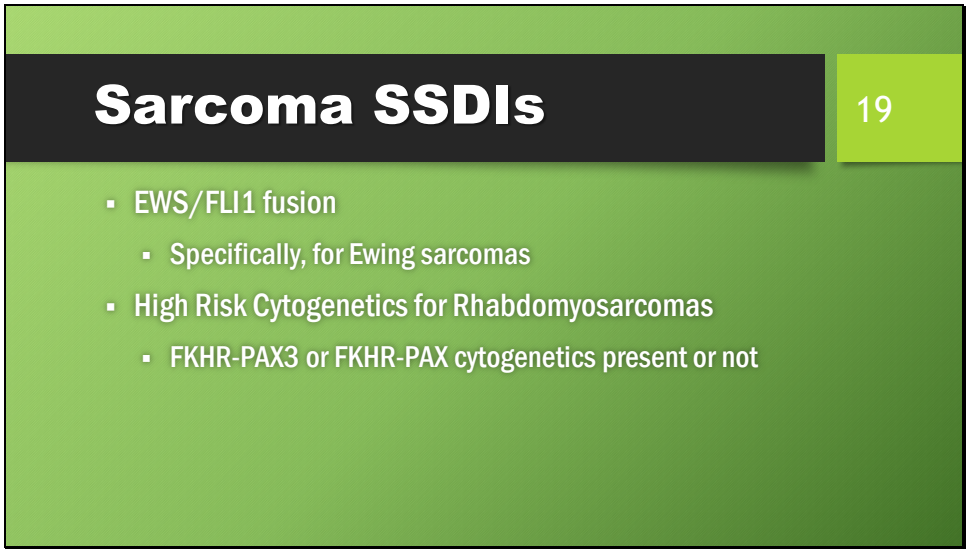
17

- International Retinoblastoma Staging System (IRSS)
  - The most current staging system for RBL.
  - In cases of bilateral disease, you will stage the MOST ADVANCED eye
  - Bilateral retinoblastoma is ALWAYS a single primary, regardless of timing.

Slide 18

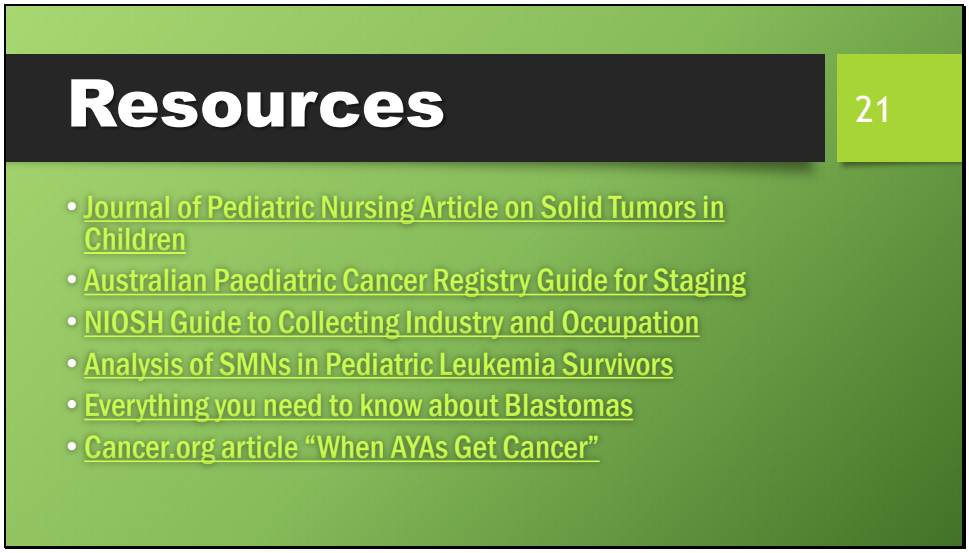

Renal SSDIs
18
National Wilms Tumor Staging System
This is one of the choices listed in the pediatric staging system field that's a part of our abstracting software.
Based on neoadjuvant chemo AND surgery
Goes from stage I-V
Bilateral disease is automatically stage V and does not require a surgical resection to be assigned this stage.
Chromosome 1p/19q LOH and 1q GOH (gain of heterozygosity) are three more prognostic indicators
1p/19q LOH are currently collected for certain brain tumors and will now be collected for certain renal tumors

Slide 19


Sarcoma SSDIs
19
EWS/FLI1 fusion
Specifically, for Ewing sarcomas
High Risk Cytogenetics for Rhabdomyosarcomas
FKHR-PAX3 or FKHR-PAX cytogenetics present or not

Slide 20

Slide 21


Resources
21
• Journal of Pediatric Nursing Article on Solid Tumors in Children
• Australian Paediatric Cancer Registry Guide for Staging
• NIOSH Guide to Collecting Industry and Occupation
• Analysis of SMNs in Pediatric Leukemia Survivors
• Everything you need to know about Blastomas
• Cancer.org article "When AYAs Get Cancer"