CBTRUS

CENTRAL BRAIN TUMOR REGISTRY OF THE UNITED STATES

# On Second Thought, Yes, We Do Have a Prevalence Estimate for Non-Malignant Brain Tumors!

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# Brain Tumors

- Controversial
  - Neuropathologists disagree often on histologic diagnosis
- Complicated
  - Over 100 histologies
- Unique
  - Rarely metastasize but many cancers metastasize to the brain site
- Orphan Disease

# Perspective that Inspired

Specimens collected by Harvey Cushing at Yale University line walls.

Neurosurgeon Harvey Cushing made the observation that some brain tumors are malignant because of their histology, and some are malignant because of their location.

# Result of Inspiration

- CBTRUS Statistical Reports 1995-2011
  - Collection of data on all brain tumors irrespective of behavior
  - Self-published
  - Dependent on volunteer CCR collaborators
- Public Law 107-260, the Benign Brain Tumor Cancer Registries Amendment Act,
  - mandates that NPCR registries collect data on all brain and CNS tumors with a behavior code of /0 (benign) and those with a behavior code of /1 (borderline), in addition to *in situ* and malignant.
- SEER, NAACCR, COC voluntarily agreed to incorporate registration of these tumors in their standard practices.
- CBTRUS Statistical Reports 2012-forward
  - data from NPCR & SEER
  - Peer-reviewed
  - Published as yearly Supplements to *Neuro-Oncology,* journal of the Society for Neuro-Oncology

# Exceptional Collection Practice

- Benign and borderline primary intracranial and CNS tumors with a behavior code of /0 or /1 in ICD-O-3 are collected for ICD-O-3 sites, C70.0-C70.1, C70.9, C71.0-C71.9, C72.0-C72.5, C72.8-C72.9, effective with cases diagnosed January 1, 2004
  - *CDC/NPCR Data Collection of Primary Central Nervous System Tumors SEER Program Coding and Staging Manual 2014*

- WHO - Pilocytic/Juvenile astrocytomas are given an uncertain behavior and grade 1
  - *2000 WHO Classification of Tumours of the Central Nervous System*

- ICD-O-3 - Pilocytic/Juvenile astrocytomas are assigned behavior code /1
  - *International Classification of Diseases for Oncology, Third Edition*

- Pilocytic/Juvenile astrocytomas are reportable as /1; code the histology and behavior as 9421/3 for continuity
  - *SEER Program Coding and Staging Manual 2014*

# Policy with Benefits

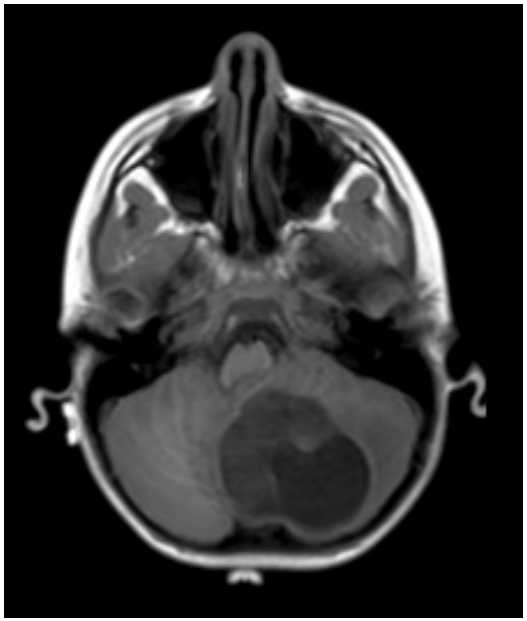
We have trends, survival and **prevalence** for a bone fide primary non-malignant brain tumor!

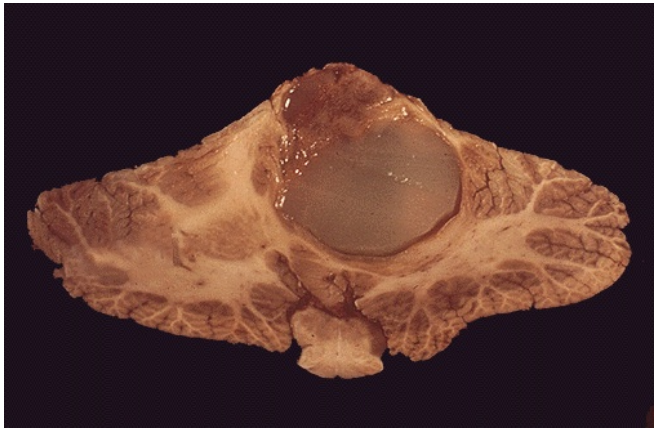
**Pilocytic Astrocytoma!**

**Cheers!**

# Snapshot of Pilocytic Astrocytoma

- Circumcised, slow growing, often cystic tumor (WHO 2007)
- Location influences presentation, treatment and survival (WHO 2007)

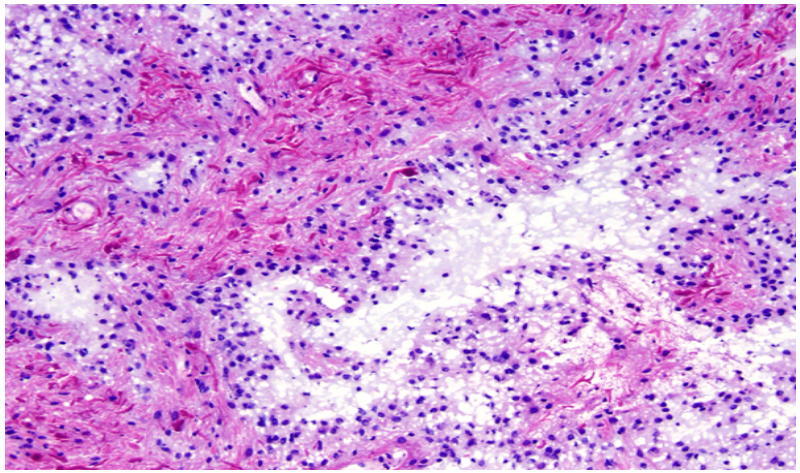




- Many can be removed totally (WHO 2007)

# Snapshot of Pilocytic Astrocytoma

- Can have an unpredictable atypical behavior even though sometimes referred to as benign (no mitosis and no necrosis) (WHO 2007 & SEER Brain Book, unpublished 2003))
  - /1 ICD-O-3 behavior



# Snapshot of Pilocytic Astrocytoma

- Included in low grade gliomas; most common glioma in children age 0-14 years (Infant & Childhood Report)
- WHO Grade I assignment is 91.8% (CBTRUS Report, 2008-2012)

| WHO GRADING SYSTEM | |
|---|---|
| **Grade I-Pilocytic astrocytoma** | Benign cytological features |
| **Grade II-Low-grade astrocytoma** | Moderate cellularity-no anaplasia or mitotic activity |
| **Grade III- Anaplastic astrocytoma** | Cellularity, anaplasia, mitoses |
| **Grade IV-Glioblastoma** | Same as Grade III plus microvascular proliferation and necrosis |

# Snapshot of Pilocytic Astrocytoma

- Most common brain tumor in children age 5-14 years; median age is 12 years (CBTRUS Report, 2008-2012)
- Principal CNS tumor of NF1 (WHO 2007)
- 33.2% of all primary brain and CNS tumors in children age 0-14 years (Infant & Childhood Report)
- Occurs equally in males and females (CBTRUS Report, 2008-2012)

- Average annual age-adjusted incidence is 0.34 per 100,000, approximately 1,010 cases diagnosed yearly (CBTRUS Report, 2008-2012)

- 10 year survival is 95.9% (age 0-19 years) (CBTRUS Report, 2008-2012)

# Prevalence

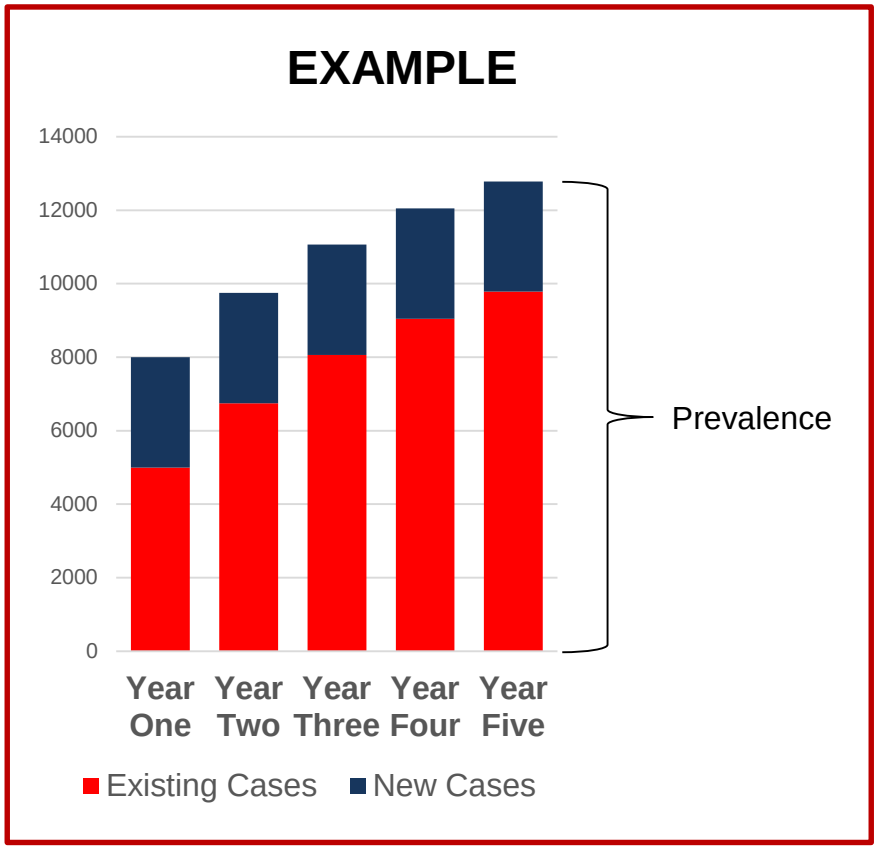
- Total Number of persons with a diagnosis at a point in time
- Relationship between incidence, survival and demographics
- Accurate estimation requires > 15 years of follow up

# Data Sources

- Used data from **three datasets**:
  - Central Brain Tumor Registry of the United States (CBTRUS) from 1995-2010
    - Largest aggregation of population-based incidence data on primary CNS tumors in the US.
    - 51 registries- 46 from National Program of Cancer Registries (NPCR), and 5 from SEER.
    - Covers ~99% of US Population.
  - Surveillance, Epidemiology and End Results (SEER)
    - SEER 9 Registries from 1975-1994
      - Covers ~ 15% of US Population.
    - SEER 18 Registries from 1995-2010
      - Covers ~ 28% of US Population.

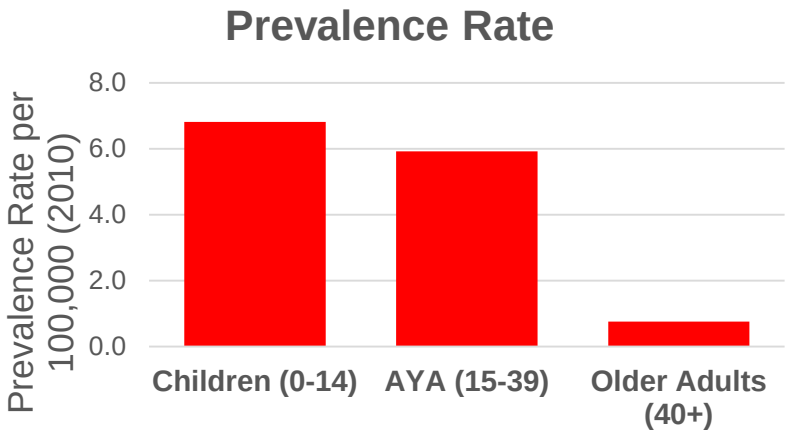
# Calculating Prevalence

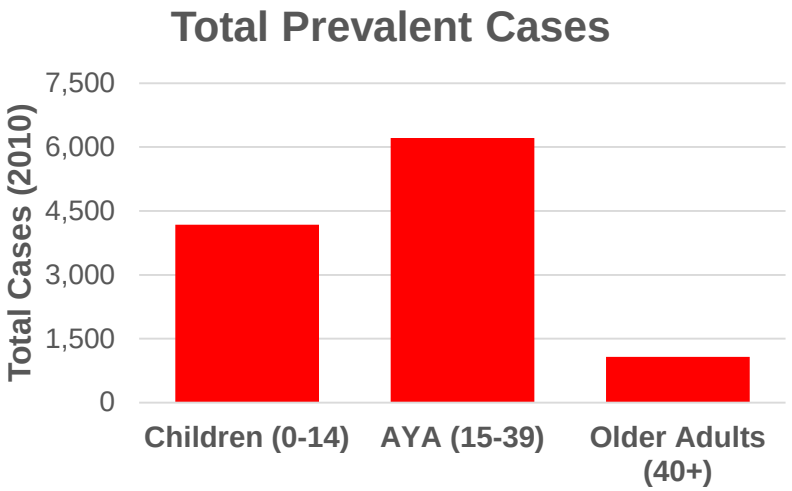
- Prevalence is a sum of **observed** and **unobserved** prevalent cases
  - Observed cases: incident cases in CBTRUS dataset
  - Unobserved cases: modeled by extrapolating from SEER 9 to entire US population
- Time as a prevalent case is estimated using SEER survival proportions
  - Assume that survival drops to zero after 15 years



# Results

- Pilocytic Astrocytoma is **most prevalent** primary brain tumor in children (age 0-14 years) and adolescents/young adults (age 15-39 years)
- Approximately 1/3 of all prevalent tumors in children



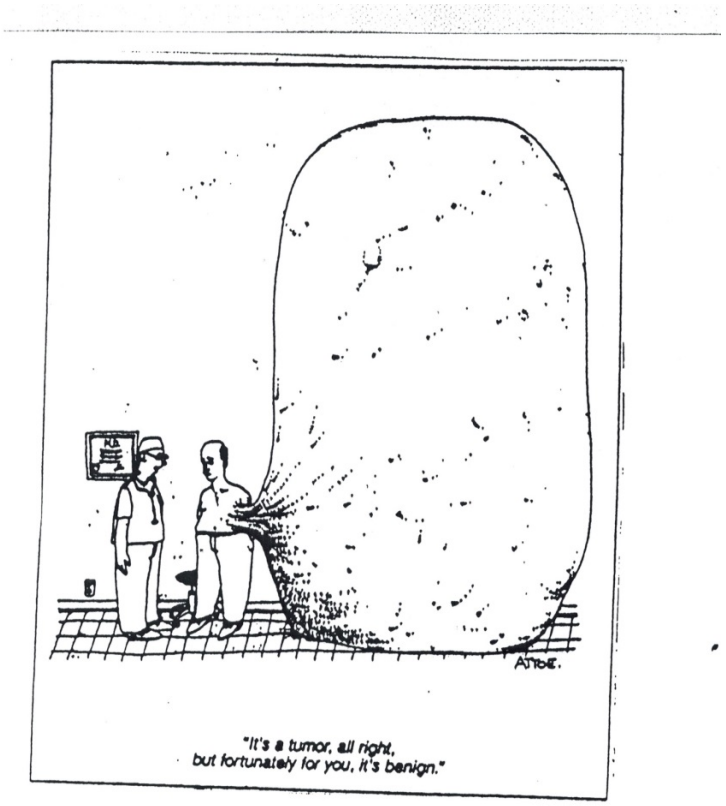
# Impact

- Pilocytic astrocytoma is one of the most common cancers in children and has relatively good survival – as a result is highly prevalent
- Actual 15 year survival for pilocytic astrocytoma is much higher than zero
  - Assumptions used to generate these calculations likely **underestimate** the impact of pilocytic astrocytoma

# Effects on Children

- All children irrespective of location have problems with sustained attention and speed
- Phenomenon of growing into deficits
- Years of Life Lived
  - Social cost
  - Financial cost
- Second primaries
  - diagnostic monitoring – CT scans

# The Reality



# Future

- Prevalence for all primary brain tumors by histology will be available.
- 2019 minimum of 15 years worth of data will be available for non-malignant brain & other central nervous system tumors, 2004-2019
- 25th CBTRUS Report!

# Acknowledgements

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