Brain tumours in young people

PROFESSOR ANNA NOWAK

MBBS FRACP PHD

MEDICAL ONCOLOGIST – NEURO-ONCOLOGIST
SCGH AND UWA

Real courage is when you know you're licked before you begin, but you begin anyway and see it through no matter what.

Harper Lee, To Kill a Mockingbird

Key messages

- Brain tumours are one of the commonest causes of cancer death in 15-25 year olds
- Treatment intent is important
  - Is treatment for palliation?
  - Or do we have a shot at cure?
- Disabilities from diagnosis
- Impact on cognition, language and emotional regulation
- Impact of medication on AYA
- Highly emotive and distressing situation for patients and families

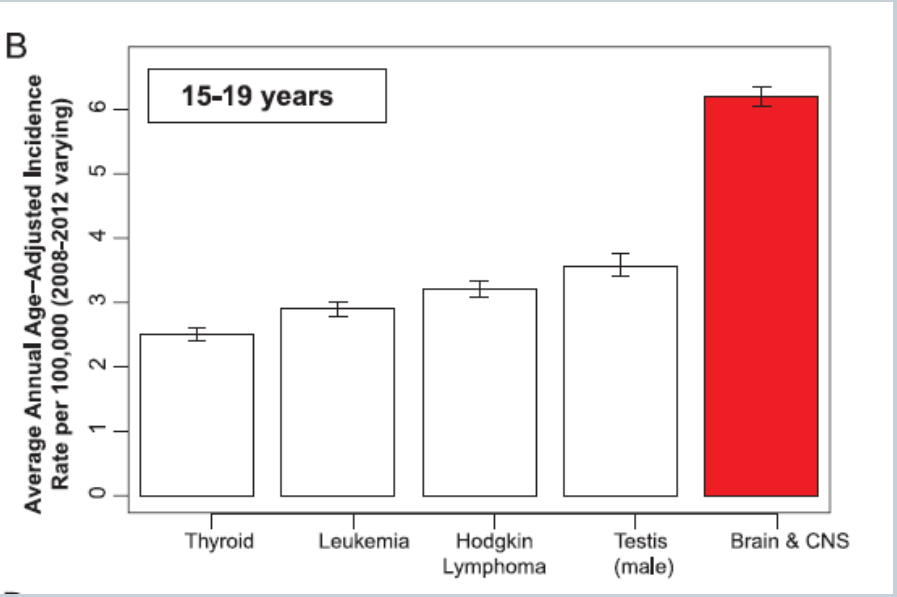
How big is this problem?

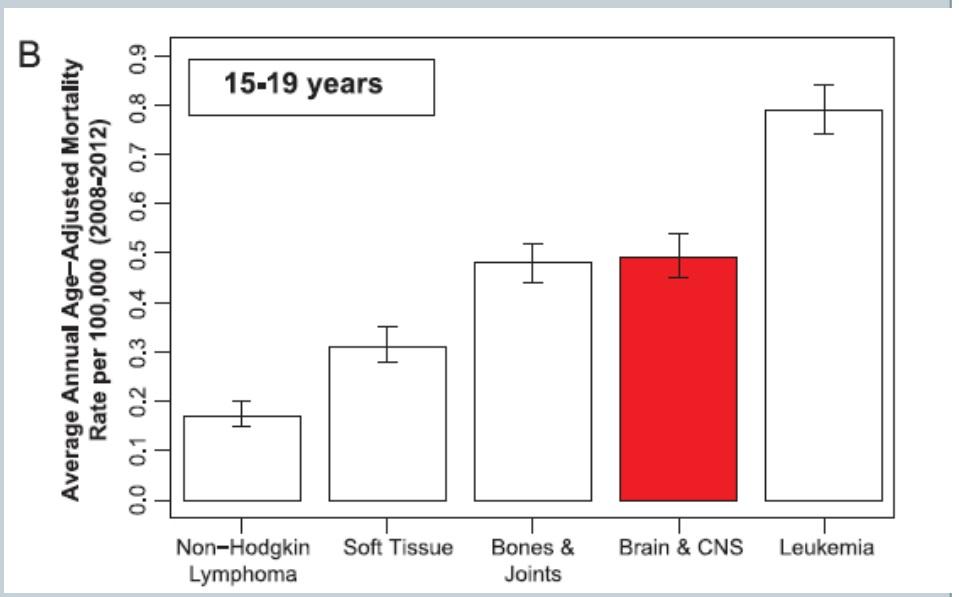
- US figures[1] – 3.3 per 100,000 (15-39 yo)
- ~ 5% of all cancers aged 15-39
- 3rd most common cause of cancer death ages 15-39

- Cancer Registry WA (2012):
  - 30 brain cancer diagnoses 15-39
  - 10 of 63 cancer deaths in this age group from brain cancer

1. Ostrom Q.T. NeuroOncology 2016

Most common cancer aged 15-19



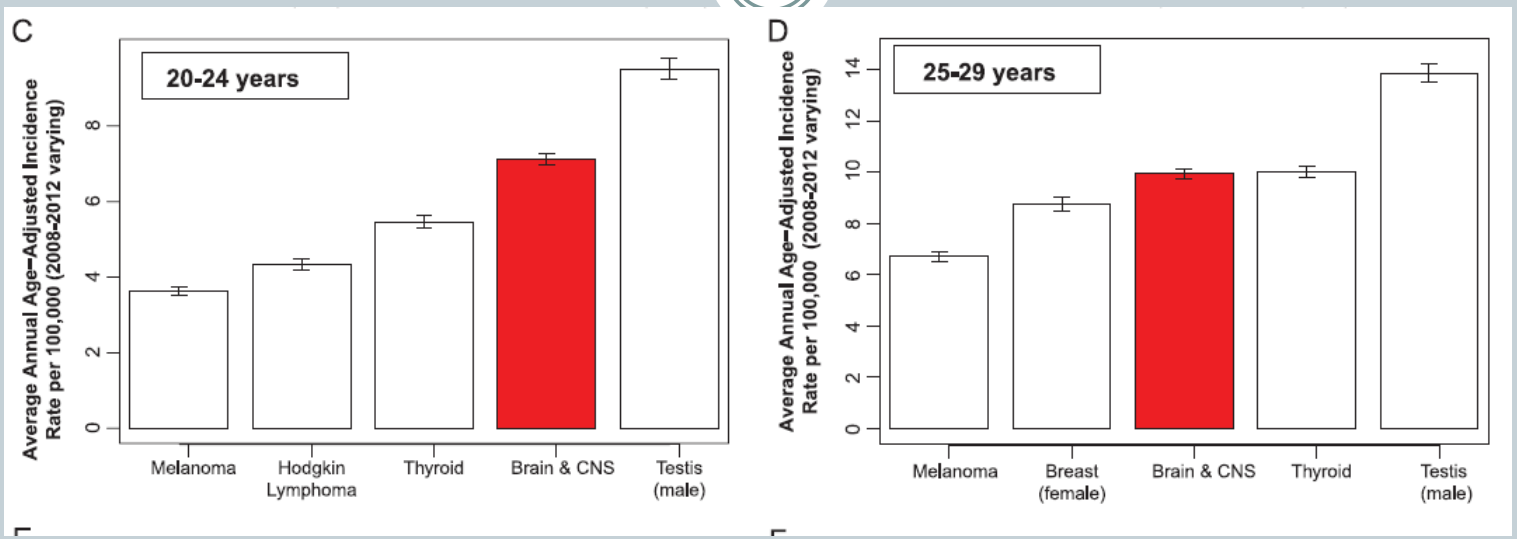


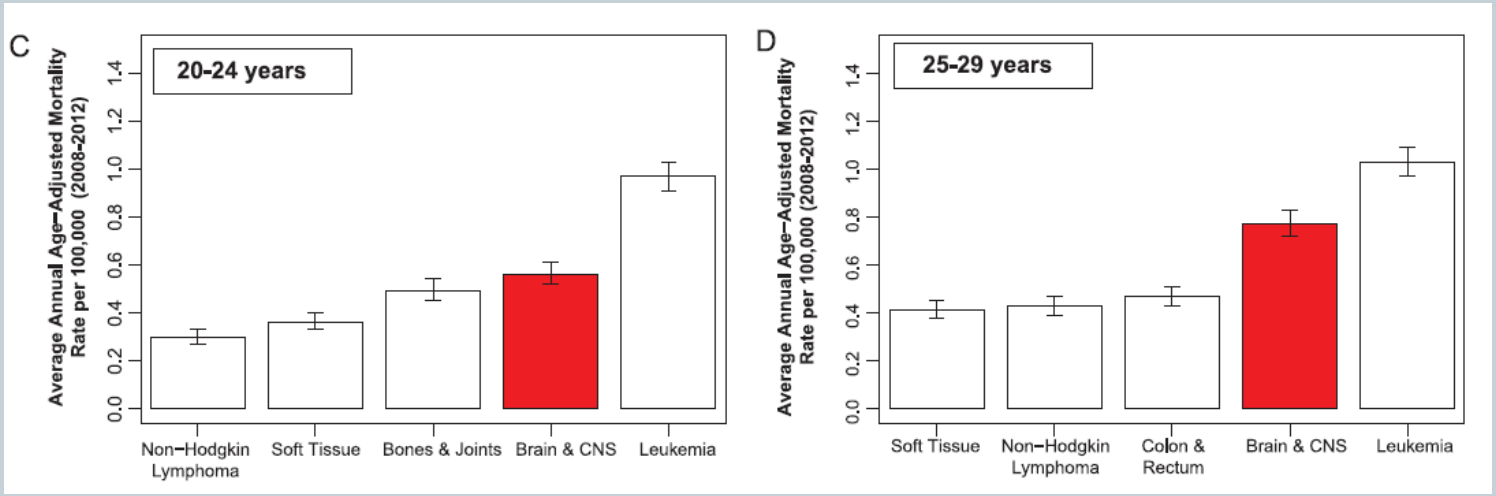
Incidence

Mortality

1. Ostrom Q.T. NeuroOncology 2016

Important cause of cancer and cancer death 20-29





1. Ostrom Q.T. NeuroOncology 2016

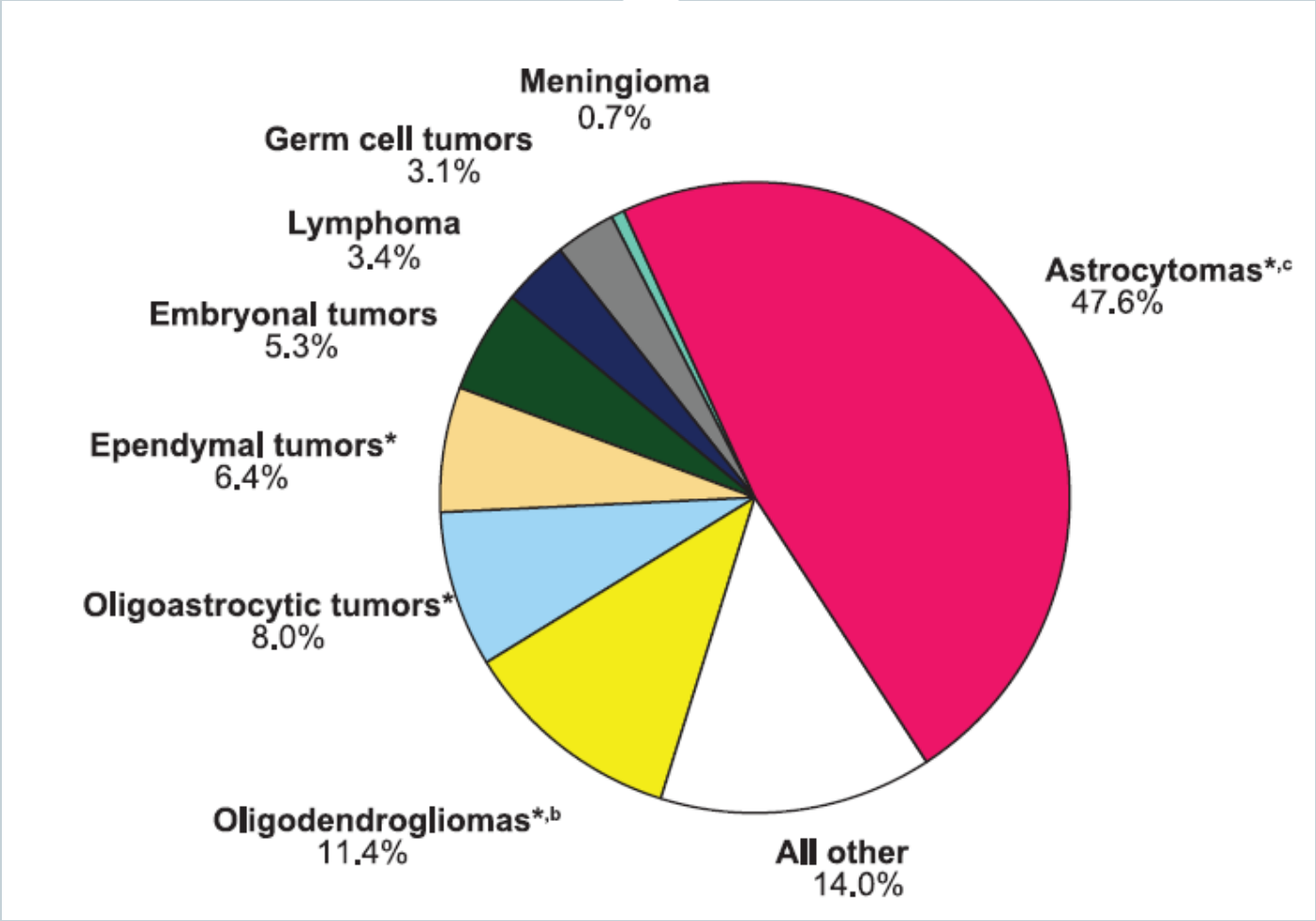
A simplified classification of primary brain tumours

- Glioma
  - Astrocytoma
  - Oligodendroglioma
  - Oligoastrocytoma
  - Ependymoma (rare)
- Adults with tumours which are usually paediatric
  - Medulloblastoma/PNET
  - Pineoblastoma
- Other rarities
  - Germ cell tumours (peak during adolescence)
  - Atypical/malignant meningioma

A simplified classification of primary brain tumours

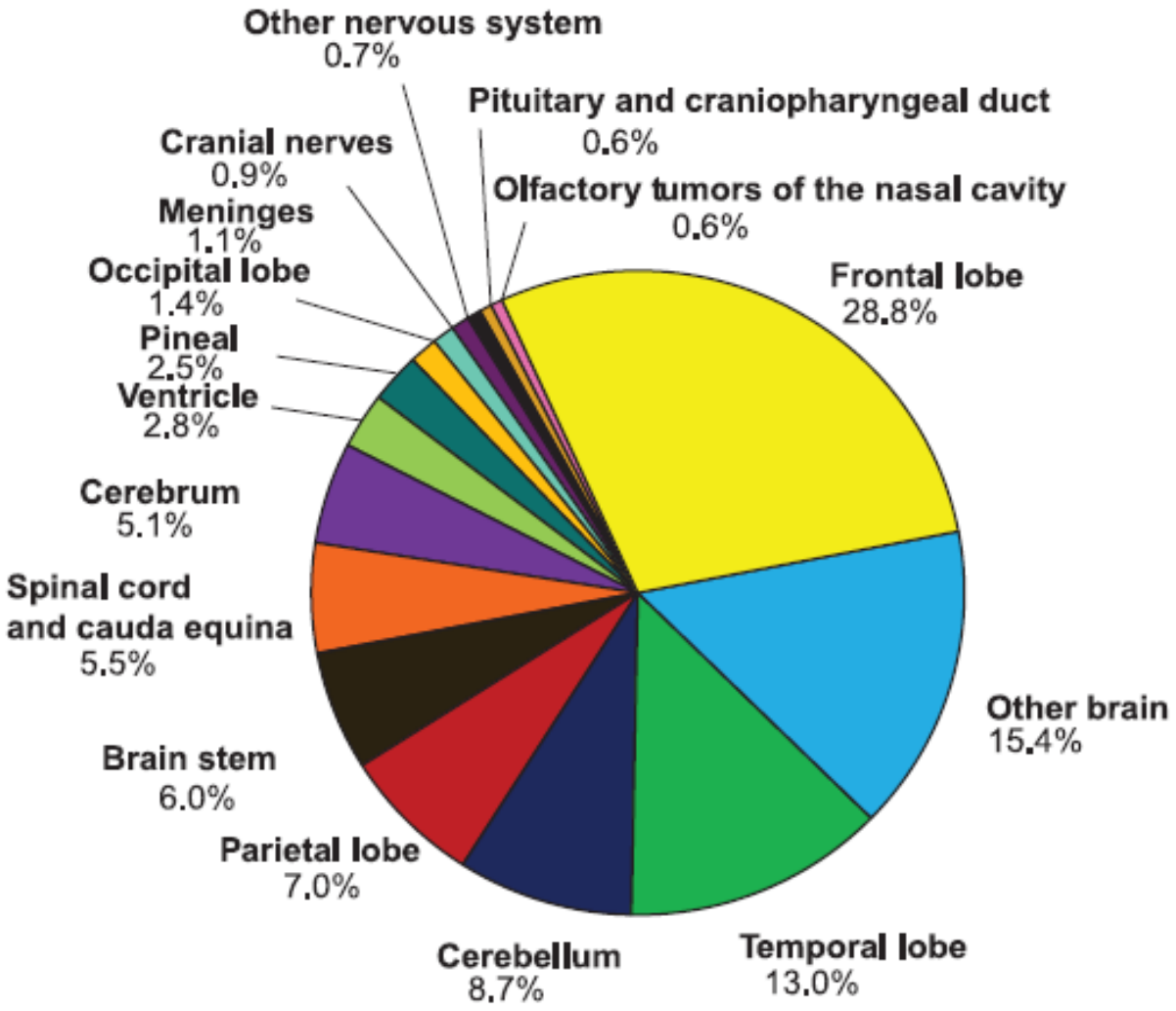
- Glioma
  - Astrocytoma
  - Oligodendroglioma
  - Oligoastrocytoma
  - Ependymoma (rare)
- Adults with tumours which are usually paediatric
  - Medulloblastoma/PNET
  - Pineoblastoma
- Other rarities
  - Germ cell tumours (peak during adolescence)
  - Atypical/malignant meningioma

Types of malignant brain tumour – young adults



1. Ostrom Q.T. NeuroOncology 2016

Malignant brain tumours – location, location, location



1. Ostrom Q.T. NeuroOncology 2016

Prognosis is variable – get the facts

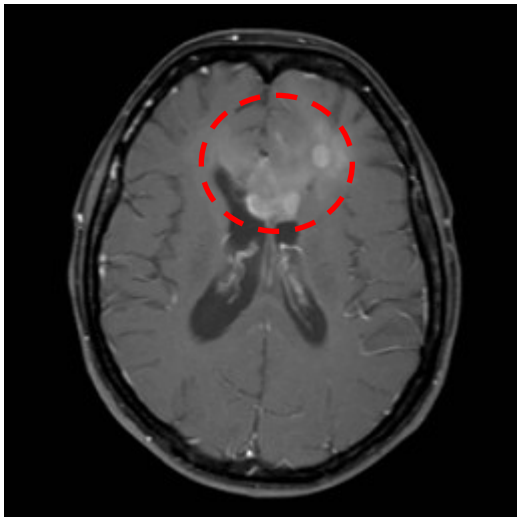
- Prognosis varies substantially with
  - Diagnosis
  - Grade of tumour
  - Molecular characteristics
  - Treatment
- BUT – loss of function varies with site, not prognosis
- SO – some functionally impaired patients may have a good prognosis; and some unimpaired patients have a poor prognosis

Treatment pathways for glial tumours

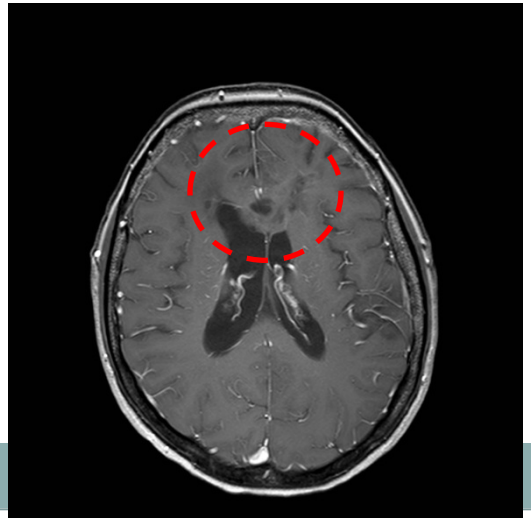
- Grade I- paediatric, surgically managed
- Grade II – slow growing, pervasive, years to diagnosis (antecedent personality change, drug and alcohol use) → surgery, radiotherapy, chemotherapy → eventual resistance, transformation and death
- Grade III – variable prognosis → surgery, radiotherapy, chemotherapy → eventual resistance, transformation and death
- Grade IV – uniform poor prognosis → surgery, chemoradiotherapy, more chemo → death

What is the goal of chemotherapy and radiotherapy?

- Chemo to enhance effects of radiotherapy
- Prolong time to progression
- Increase survival
- Decrease tumour burden
- Improve symptoms by decreasing tumour burden



Before

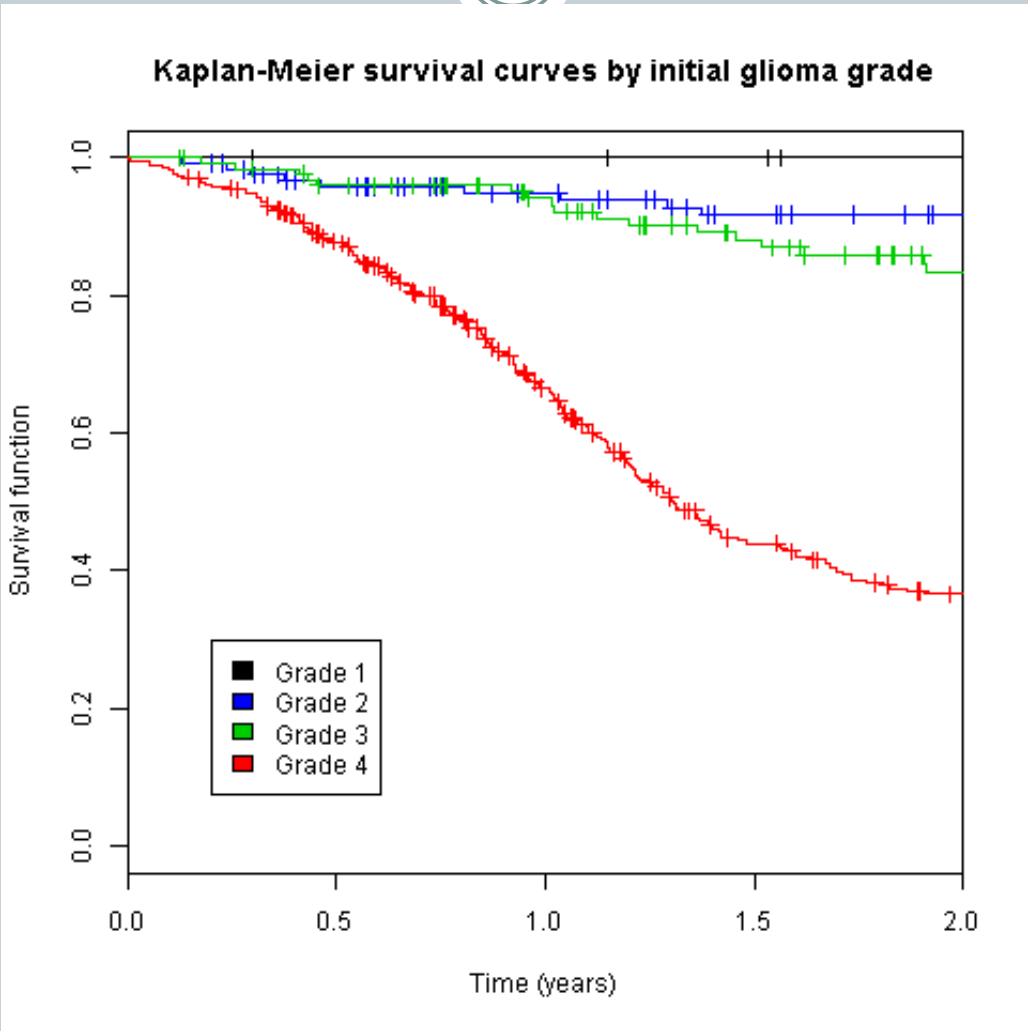


After

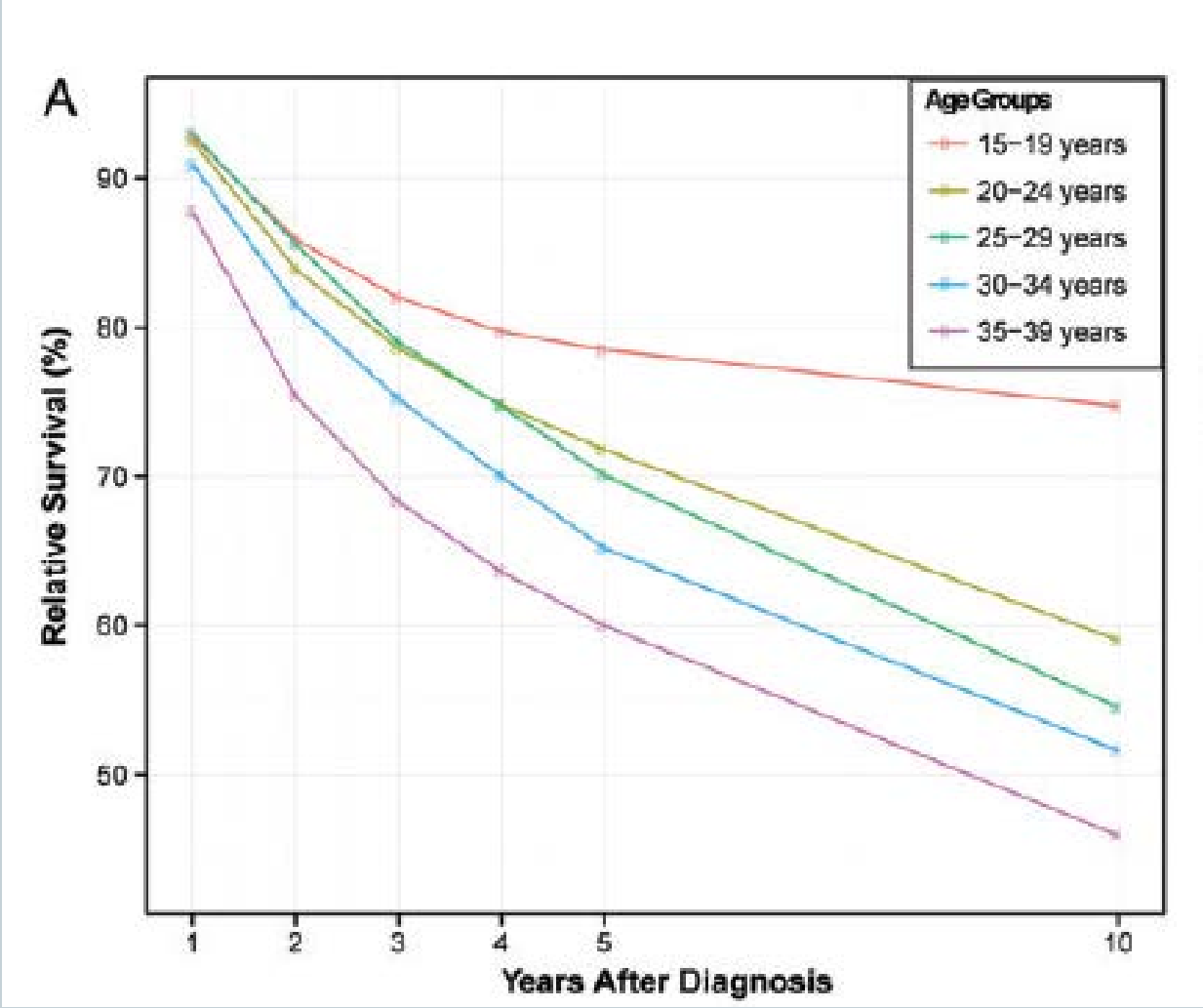
Treatment - most of the 'rarities'

- Treatment goal is cure – but only achieved in a proportion
- Aggressive surgery
- Radiotherapy (often craniospinal)
- Aggressive multiagent chemotherapy – 6 months +
- May include stem cell transplant (at relapse)
- High treatment morbidity
- Paediatric vs. Adult regimens and chemo tolerance
- Little evidence-based treatment → extrapolated from kids

Survival by grade - glioma



Younger age = better prognosis for glioma



1. Ostrom Q.T. NeuroOncology 2016

Acquired disabilities

- Frontal lobe syndrome
  - Judgement, personality change, motivation, behaviour, social appropriateness
  - Antecedent drug use – long history of poor judgement
- Hemiparesis and gait disturbance
- Seizures
- Expressive dysphasia
- Cognitive disabilities, executive dysfunction

AYA brain tumours and independence

- Inability to drive
- Loss of income and financial independence
- FIFO + brain tumour = no job
- Loss of ability to live independently (eg. shared accomodation)

- Parents →
  - Driver
  - Financial support
  - Moving back home
  - ADD parental distress, protectiveness and concern

AYA brain tumours affect normal young adult experiences

- Loss of immature or new relationships
- Functional or cognitive difficulties and study/workplace
- Alcohol contraindicated
- Future fertility – effect of treatments (and tumour)
- Sexuality - reduced libido
- International travel – no longer possible
- Feelings of futility about study and training
- Friends and acquaintances- what to say?

AYA brain tumours affect physical appearance

- Patchy hair loss at radiotherapy entry sites
- Corticosteroids = changes in appearance, weight gain
- Gait disturbance and use of walking aids

Effects of supportive care treatments

- Anticonvulsants
  - Drowsiness
  - Cognitive change
  - Irritability and aggressiveness (levetiracetam)
  - Weight gain, hirsutism
  - Alcohol contraindicated
- Dexamethasone
  - Weight gain, facial change
  - Acne
  - Insomnia
  - Irritability and hypomania
  - Proximal myopathy

Impact of frontal lobe syndromes, cognition and language on medical care

- Those with frontal lobe syndrome may struggle with executive functions, timekeeping, attendance, and motivation – notorious for not turning up!
  - Give a second chance, enlist help (family, admin staff) to prompt attendance
- Those with cognitive dysfunction may have poor recall and understanding
- Receptive or expressive dysphasia – communication difficulties

Challenges of caring for dying young adults

- Heightened individual and family distress
- Time needed +++
- Seeking alternatives
  - Complementary and alternative medicine
  - Second/third/fourth opinions
  - International opinions – both legitimate and maverick
- Requests for ongoing futile or unproven therapy
- Ensuring patient has a voice, amongst cognitive and physical changes

Memorable challenges

- Young ex-prisoner (drugs) with severe headaches from huge low grade glioma
  - Could not find a GP to prescribe opiates (which he needed in high doses due to tolerance)
- Young man with ?cured medulloblastoma whose mother has become over protective
- Young woman with ?cured ependymoma who is struggling to relinquish the sick role which made her feel special
- Young scientist with long term survival from grade III glioma unable to accept diminished roles in workplace 5+ years after treatment
- Highly intelligent uni student who won't tell anyone other than his mother that he has a grade III astrocytoma for fear of being treated differently

Thank you

QUESTIONS WELCOMED