

# **Diagnosing brain tumours in children - clinical aspects**

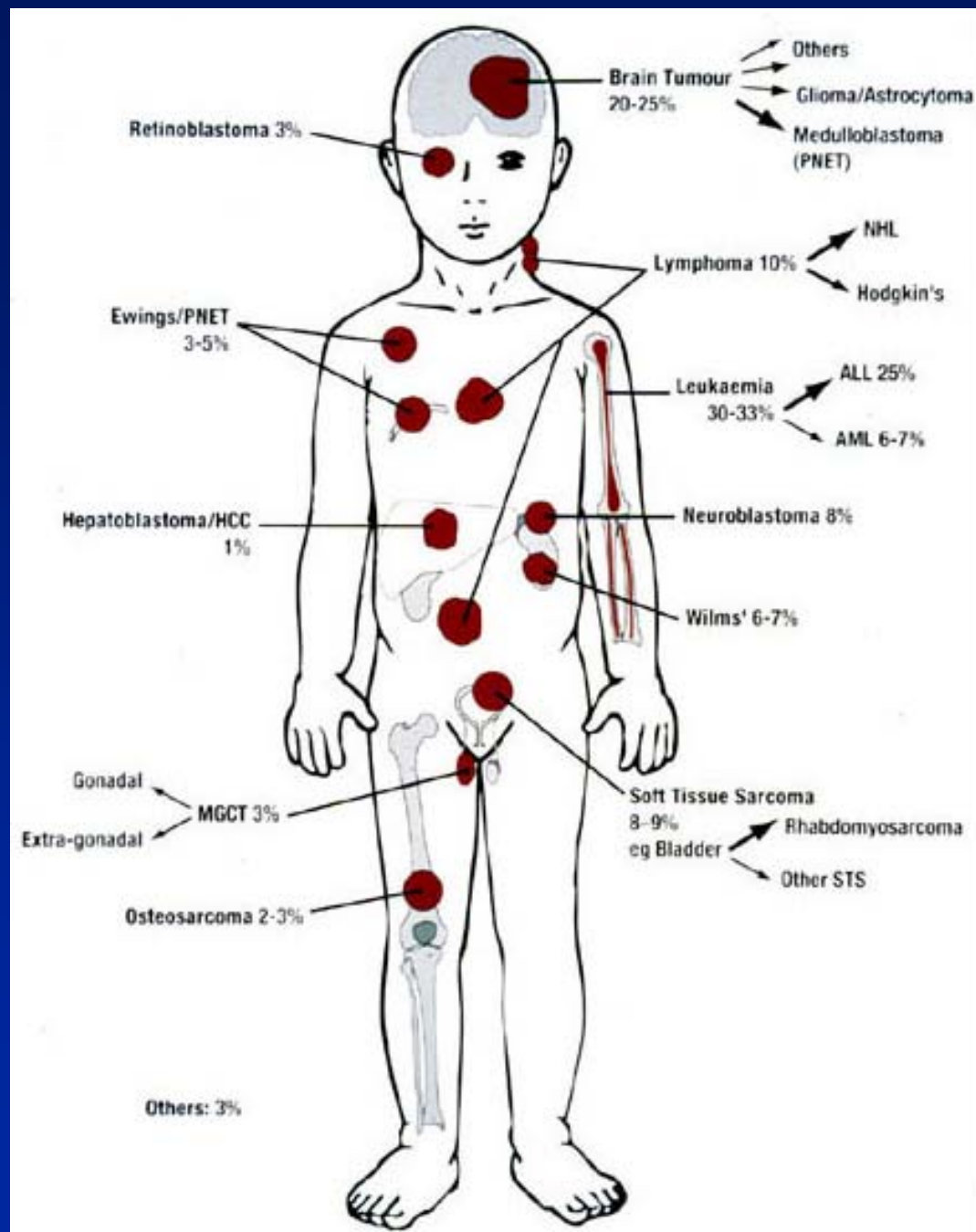
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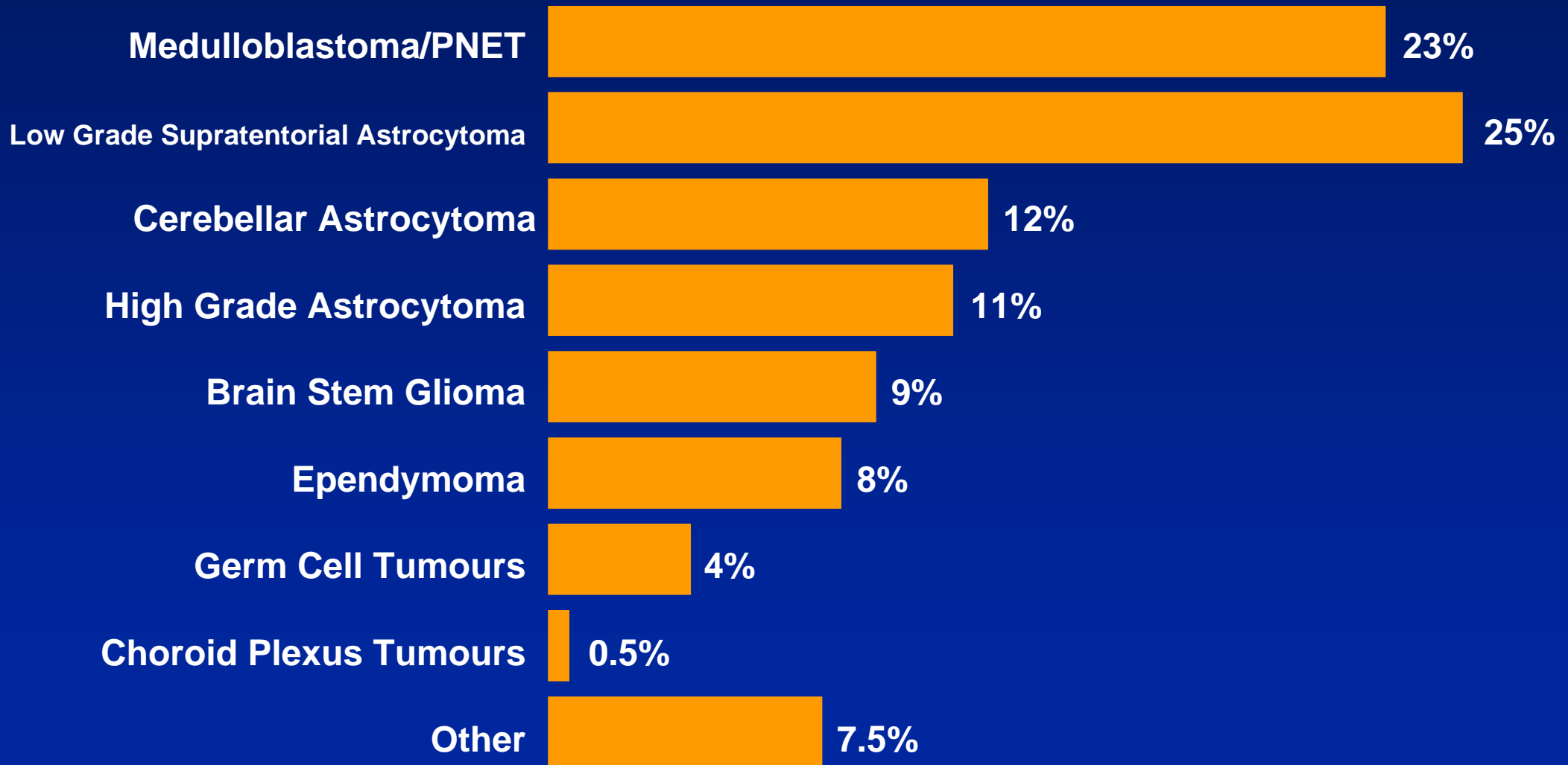
**September 2011**

# Childhood Cancer: Types and Frequencies 0-14 years

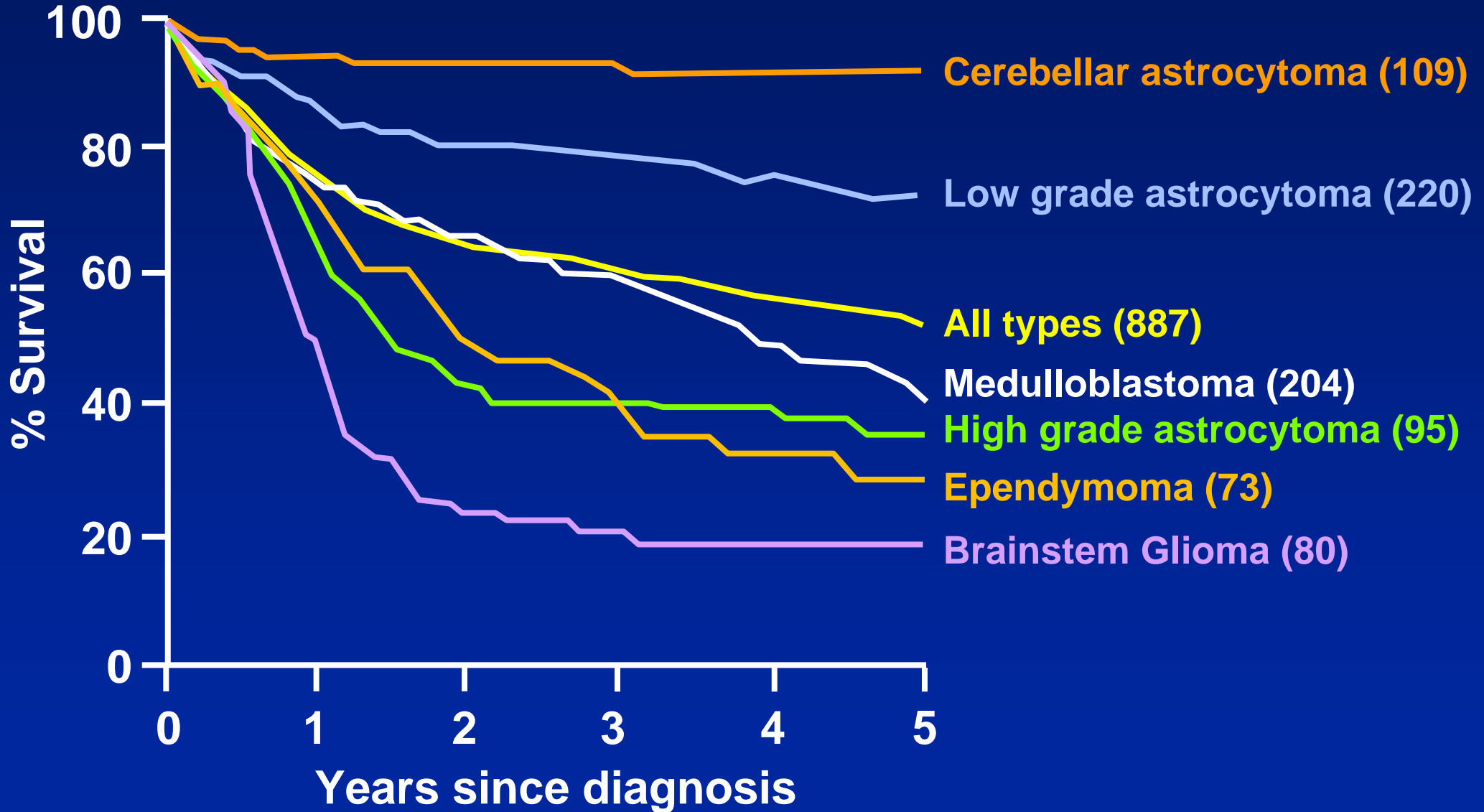
Between  
1300-1400 new  
UK cases every year



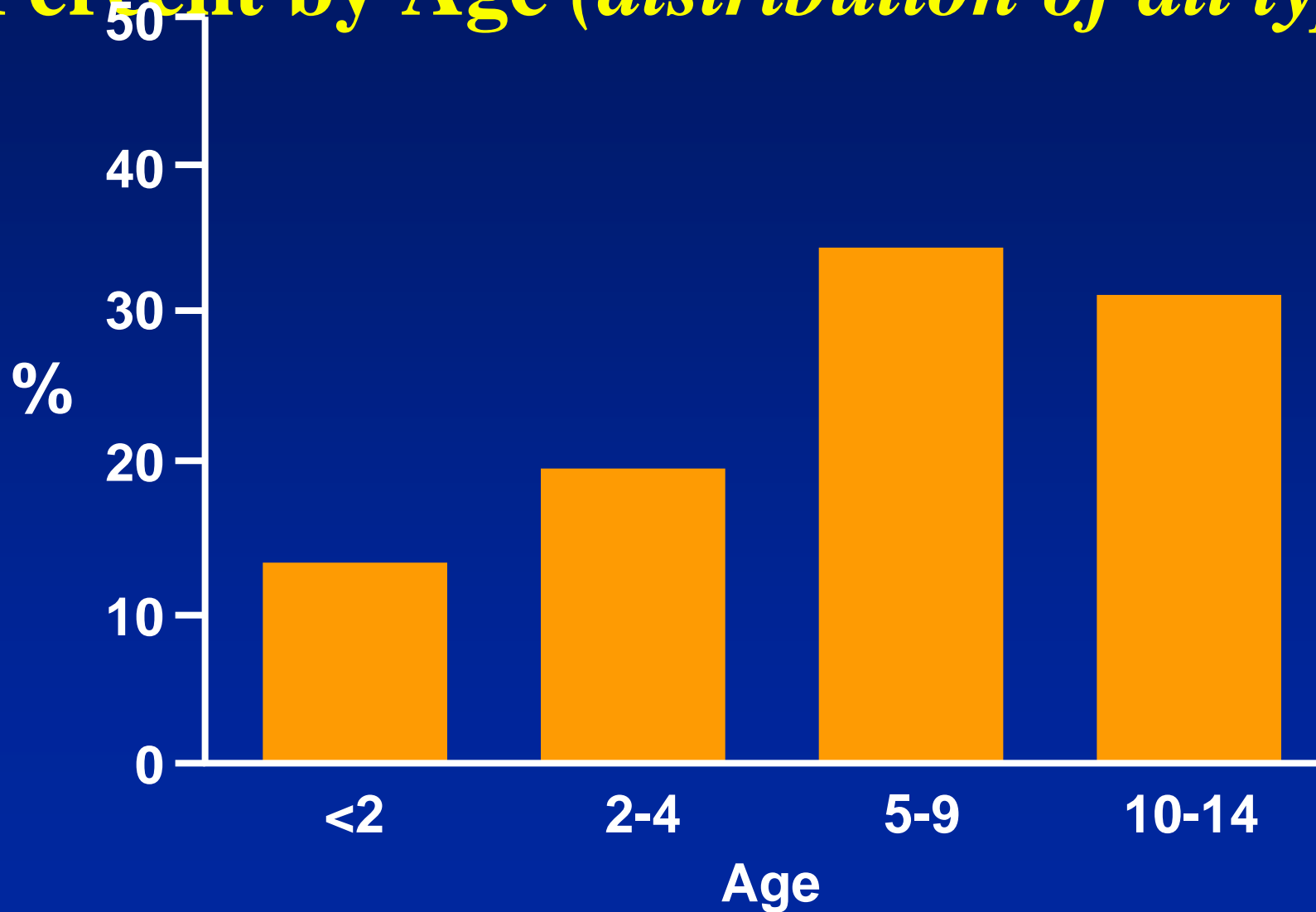
# Malignant Brain Tumours in Children <15 years: Distributed by tumour type



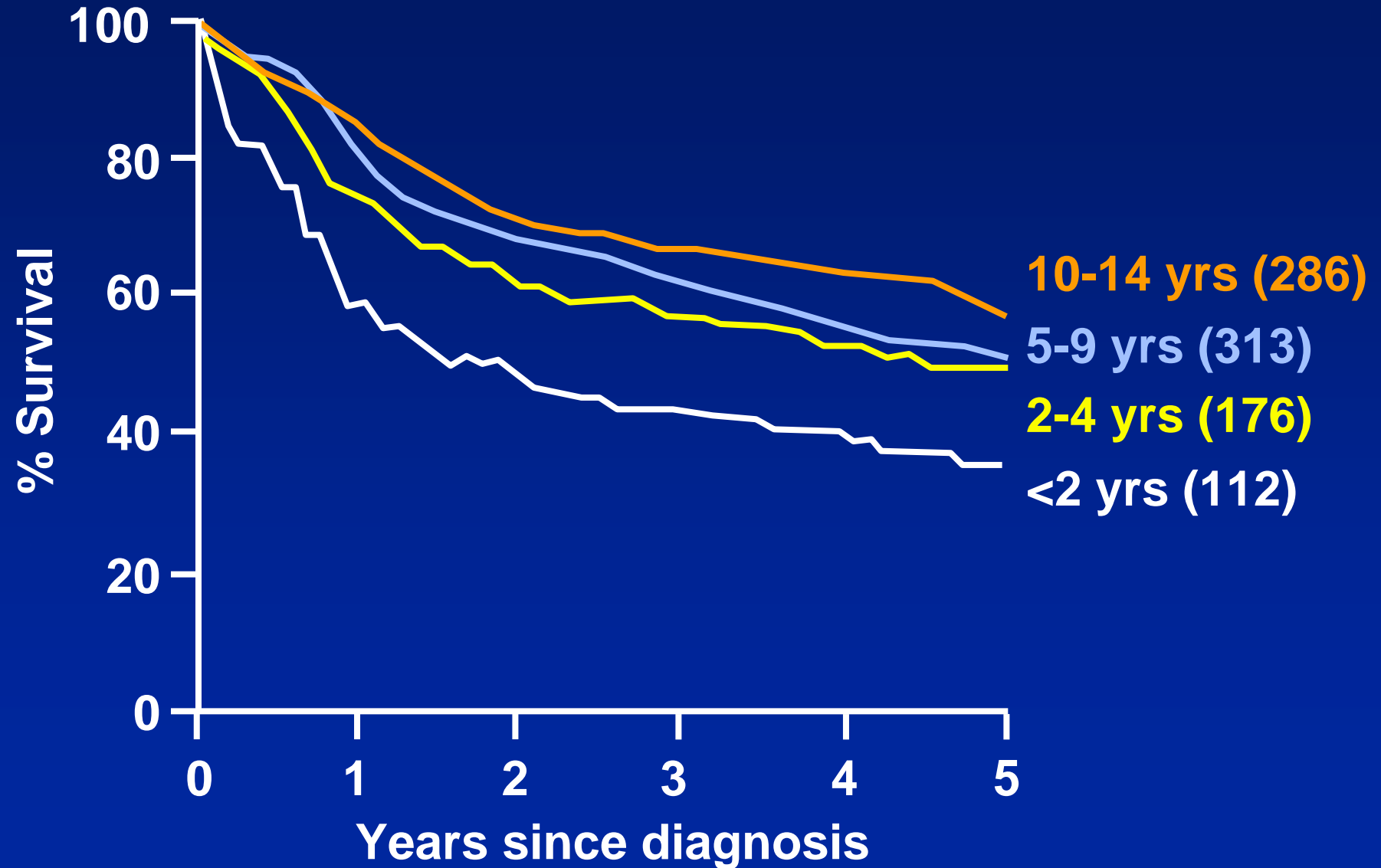
# Survival According to Tumour Type



# Percent by Age (*distribution of all types*)



# Survival According to Age at Diagnosis



# How do children with brain tumours present?

- Presentation of childhood CNS tumours: a systematic review and meta-analysis
- The Lancet Oncology, [Volume 8, Issue 8, Pages 685 - 695, August 2007](#)
- doi:10.1016/S1470-2045(07)70207-7  
Published Online: 18 July 2007
- *Wilne S, Collier J, Kennedy C, Koller K, Grundy R, Walker D.*

# Findings of meta-analysis

- ‘Apart from raised intracranial pressure, motor and visual system abnormalities, weight loss, macrocephaly, growth failure, and precocious puberty also suggest presence of an intracranial tumour. Children with signs and symptoms that could result from a CNS tumour need a thorough visual and motor system examination and an assessment of growth and pubertal status. Occurrence of multiple symptoms and signs should alert clinicians to possible CNS tumours.’
- *Wilne S, Collier J, Kennedy C, Koller K, Grundy R, Walker D.*

# Symptoms and signs

- **Mass effect and/or hydrocephalus**
- **Affected bit of brain not doing what it should do (loss of function)**
- **Affected bit of brain doing what it should not do (seizures)**

# CNS tumours present late

- Median time from first symptom to diagnosis is:
  - 1 - 3 weeks Wilms tumour
  - 3 - 7 weeks ALL
  - 9 weeks posterior fossa tumours
  - 19 weeks for hypothalamic tumours
  - 14 weeks for all brain tumours

*Saha et al 1993 Arch Dis Child 68 771-4*

*Pollock et al 1991 J Pediatr 119 725-32*



# Guide to assist in early diagnosis

- *Arch Dis Child* doi:10.1136/adc.2009.162057
- Original article
- The diagnosis of brain tumours in children: a guideline to assist healthcare professionals in the assessment of children who may have a brain tumour
- *Wilne S, Collier J, Kennedy C, Koller K, Grundy R, Walker D.*

# The Diagnosis of Brain Tumours in Children: A Guideline for Healthcare Professionals

## HEADACHES:

- Consider a brain tumour in any child presenting with a new, persistent\* headache
- Brain tumour headaches occur at any time.
- Children aged younger than 4 years may not be able to complain of a headache—observe behaviour.

## CNS IMAGING REQUIRED WITH:

- Persistent\* headaches that wake a child from sleep
- Persistent\* headaches that occur on waking
- Persistent\* headaches at any time in a child younger than 4 yrs
- Confusion or disorientation and a headache

## COMMON HEADACHE PITFALLS:

- Failure to re-assess a child with migraine or tension headache when the headache character changes

\* Persistent = continuous or recurrent headache present for more than 4 weeks

## NAUSEA AND VOMITING:

- Consider a brain tumour in any child with persistent\* nausea and / or vomiting.
- A child with persistent\* nausea and / or vomiting requires specialist assessment within 2 weeks

## CNS IMAGING REQUIRED WITH:

- Persistent vomiting on awakening (NB: exclude pregnancy where appropriate)

## COMMON VOMITING PITFALLS:

- Failing to consider a CNS cause for persistent nausea and vomiting

\* Persistent = nausea and / or vomiting present for more than 2 weeks

## VISUAL SYMPTOMS AND SIGNS:

- Consider a brain tumour in any child presenting with a persisting\* visual abnormality
- Visual assessment requires assessment of:
  - Acuity
  - Eye movements
  - Pupil responses
  - Optic disc appearance
  - Visual fields (>= 5 yrs)
- Pre-school and uncooperative children should be assessed by hospital eye service within 2 weeks of referral.

## CNS IMAGING REQUIRED WITH:

- Papilloedema
- Optic atrophy
- New onset nystagmus
- Reduction in acuity not due to refractive error
- Visual field reduction
- Proptosis
- New onset paralytic (non-comitant) squint

## COMMON VISUAL PITFALLS:

- Failure to fully assess vision in a young or un-cooperative child—REFER IF NECESSARY
- Failure of communication between community optometry and primary and secondary care

\* Persistent = visual abnormality present for more than 2 weeks

## REFERRAL FROM PRIMARY CARE:

High risk of tumour—same day referral to secondary care

Lower\* risk—specialist assessment within 2 weeks

## IMAGING:

High risk of tumour—urgent CNS imaging

Lower\* risk—CNS imaging within 4 weeks

\* Lower risk = CNS tumour in differential diagnosis, low index of suspicion

## CONSIDER A BRAIN TUMOUR IN ANY CHILD PRESENTING WITH:

Headache

Nausea and / or vomiting

Visual symptoms and signs

reduced visual acuity and / or fields  
abnormal eye movements  
abnormal fundoscopy

Motor symptoms and signs

abnormal gait  
abnormal coordination  
focal motor weakness

Growth and developmental abnormalities

growth failure (weight / height)  
delayed, arrested or precocious puberty

Behavioural change

Diabetes insipidus

Seizures (see [www.nice.org.uk/CG020](http://www.nice.org.uk/CG020))

Altered consciousness (see [www.nottingham.ac.uk/paediatric-guideline](http://www.nottingham.ac.uk/paediatric-guideline))



## ASSESS THESE CHILDREN WITH:

History : Associated symptoms  
Any predisposing factors

Assessment of:

Visual system  
Motor system  
Height and weight  
Head circumference (< 2 yrs)  
Pubertal status

## ASSESSMENT PITFALLS:

- The initial symptoms of a brain tumour frequently mimic those that occur with common childhood conditions
- Symptoms frequently fluctuate—resolution and then recurrence does not exclude a brain tumour
- A normal neurological examination does not exclude a brain tumour
- Language difficulties—use Interpreting services if necessary

## MOTOR SYMPTOMS AND SIGNS:

- Consider a brain tumour in any child presenting with a persisting motor abnormality
- Motor assessment requires observation of:
  - Sitting and crawling in infants
  - Walking and running
  - Handling of small objects
  - Handwriting in school age children
- Brain tumours may cause a deterioration or change in motor skills—this can be subtle e.g. change in hand preference

## CNS IMAGING REQUIRED WITH:

- Regression in motor skills
- Focal motor weakness
- Abnormal gait and / or co-ordination (unless local cause)
- Bells palsy with no improvement within 4 weeks
- Swallowing difficulties (unless local cause)

## COMMON MOTOR PITFALLS:

- Attributing the abnormal balance or gait caused by a cerebellar lesion to middle ear disease
- Failure to identify swallowing difficulties and aspiration as the cause of recurrent chest infections

\* Persistent = motor abnormality present for more than 2 weeks

## GROWTH AND DEVELOPMENT:

- Consider a brain tumour in any child presenting with any combination of growth failure, delayed / arrested puberty and polyuria / polydipsia
- Early assessment is required for a child presenting with
  - Precocious puberty
  - Delayed or arrested puberty
  - Growth failure

## COMMON GROWTH AND DEVELOPMENT PITFALLS:

- Failure to consider a CNS cause in children with vomiting and weight loss
- Failure to consider diabetes insipidus in children with polyuria and polydipsia

## BEHAVIOUR:

- Lethargy is the commonest behavioural abnormality that occurs with brain tumours

## COMMON PREDISPOSING FACTORS:

- Personal or family history of a brain tumour, leukaemia, sarcoma or early onset breast cancer
- Prior therapeutic CNS irradiation
- Neurofibromatosis ([www.nfauk.org](http://www.nfauk.org))
- Tuberous sclerosis ([www.tuberous-sclerosis.org](http://www.tuberous-sclerosis.org))
- Other familial genetic syndromes



This guideline was developed by The Children's Brain Tumour Research Centre, University of Nottingham  
Funding was provided by the Big Lottery Fund in conjunction with The Samantha Dickson Brain Tumour Trust



The University of Nottingham

# Brain Pathways: Promoting Earlier Diagnosis of Brain Tumours in Children

- **Reduce** the UK median interval from symptom onset to diagnosis to less than five weeks
- **Enhance** public and practitioner awareness of symptomatology of brain and spinal tumours in children
- **Launching** a national awareness campaign in the media
- **Developing** a decision-support website aimed at professionals and the general public, to support physicians and families in selecting children for referral
- **Evaluating** the impact of this programme upon physician and public awareness of brain tumour risk
- **Measuring** the interval between symptom onset and diagnosis for all UK cases
- **Planning** further development of the programme

# Hydrocephalus



- Symptoms?
- Signs?
- How do you measure severity?
- Emergency therapy?
- How would you treat pain?

# Hydrocephalus



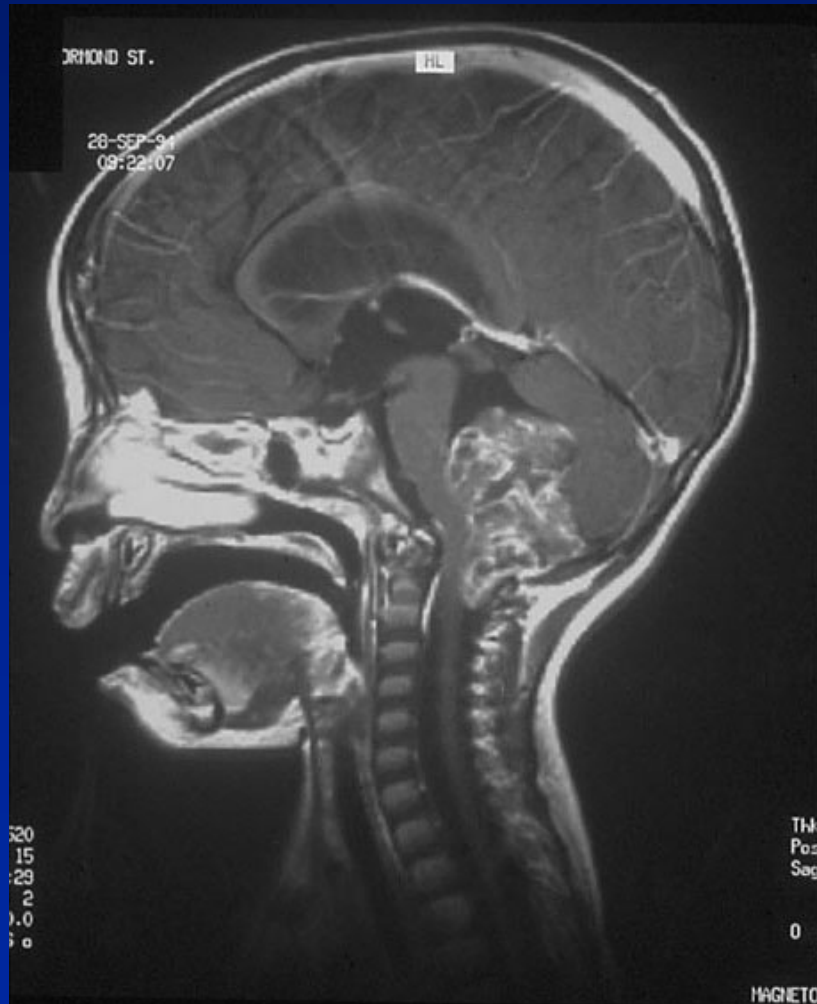
- Symptoms mimic common childhood diseases
- Often fluctuates
- Papilloedema late sign
- Glasgow coma scale
- Emergency therapy - high dose steroids and early referral to neurosurgeons
- Avoid opiates

# Posterior fossa tumours



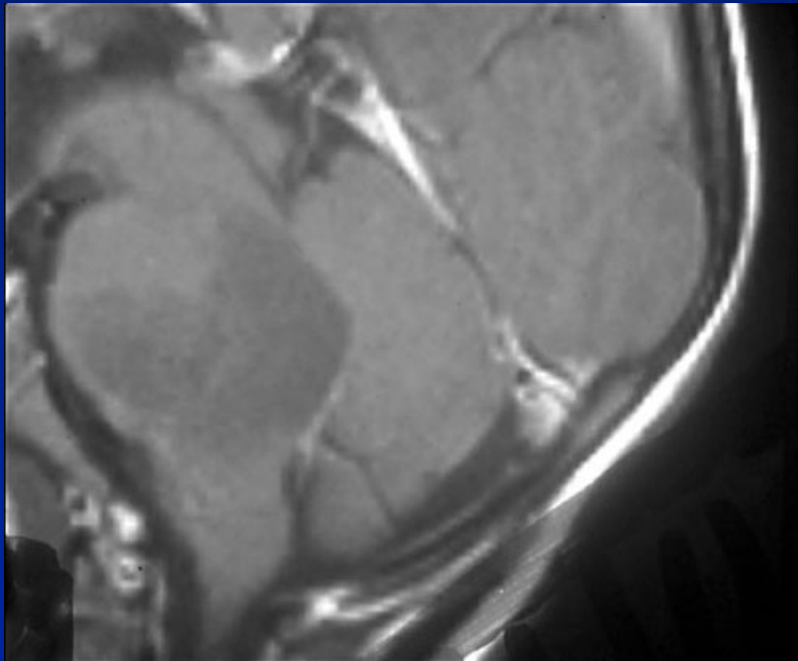
- How may they present?
- What 'areas' of the brain may be affected?
- Diagnoses?

# Posterior fossa tumours



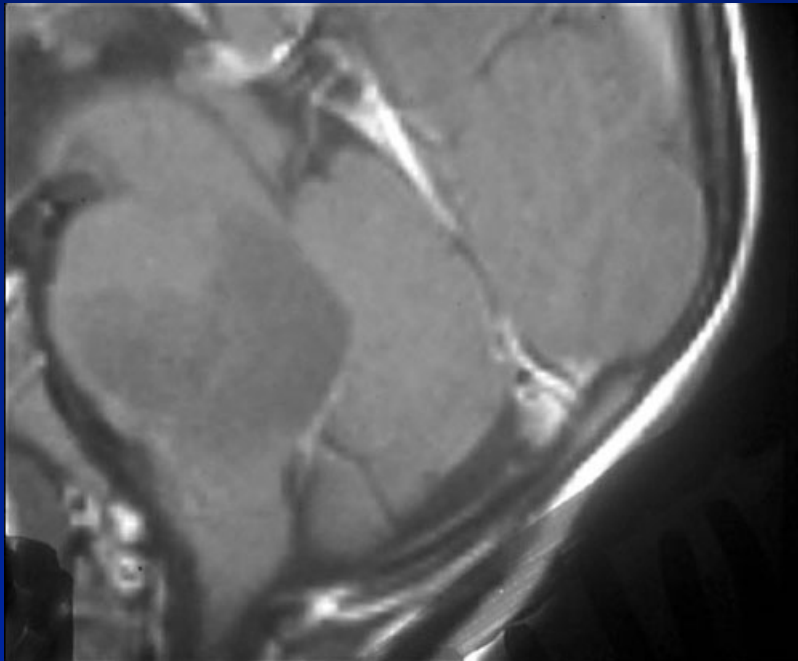
- Hydrocephalus
- Cerebellar symptoms
- Brain stem symptoms
- Diagnoses:
  - Astrocytoma
  - PNET
  - Ependymoma

# Diffuse intrinsic pontine glioma



- How may this present?
- What is prognosis?

# Diffuse intrinsic pontine glioma



- Mood changes
- Bulbar problems
- Cranial nerve signs
- Long tract signs
- Median time to recurrence 9 months
- Overall survival <10% @ 2 years

# Diencephalic syndrome



- What is going on?
- Eye signs?
- Any other symptoms or signs?
- What tumour causes this and where is it?

# Diencephalic syndrome



- Weight loss in spite of normal calorie input
- Often have nystagmus as sign of poor vision
- May have electrolyte disturbances and other problems with hypothalamic function
- Hypthalamic chiasmatic astrocytoma

# Optic tract tumours



- What is diagnosis?
- What skin changes may you find?
- Would you biopsy this?
- What assessments would you do?
- Therapy?

# Optic tract tumours



- Low grade astrocytoma
- Café au lait spots, neurofibromas
- No biopsy
- Full ophthalmology assessment
- Follow up for NF1 as well as tumour

# Summary

- High index of suspicion
- New eye signs are a worry and should never be ignored
- Scans are not curative
- Work with neurosurgeons



# **Thanks to:**

**Dr Anthony Michalski**

**Consultant Paediatric Oncologist at GOSH**

**Samantha Dickson Brain Tumour Trust**

**HeadSmart**