Paediatric Oncology: Past, present and future

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I believe that there is merit in looking at the past in order to have a perspective of accomplishments, challenges, trends and future needs. Moreover, I think that it can give us courage and determination to pursue our ideas and ideals.

R Hilkemeyer (1982)
Ancient history

- Hippocrates 460-370BC
  - First to describe a cancer - (Karakinos or crab)
  - Mentioned neck growths in children although not clear these were cancers
  - The 4 humors
    - Blood
    - Phlegm
    - Yellow bile
    - Black bile
Early descriptions

1809 – Wardrop
   – Described 24 cases of eye malignancy

1876 – CJ Duzan
   – Published a table of 182 paediatric malignancies

1921 – James Ewing (pathologist)
   – Objected to cancer rates being clumped under 30 yrs of age
Children are miniature adults?

NOT

16\textsuperscript{th} c – France children admitted to hospital
1802 – l’Hospital des enfants Malades
1852 – Great Ormond St Hospital
1955 – Children’s Hospital of Philadelphia
1962 – St Jude’s Memphis
1938 – “death in acute leukaemia in a matter of days or weeks”
1946 – “because the disease is invariably fatal treatment is aimed at modifying symptoms”
1954 – “generally fatal but with the administration of antimetabolites 25-50% can achieve remission
1959 – “with supportive anti-leukaemic therapy life can be prolonged for six months or a year
1975 – “substantial numbers of children with ALL are in remission 3 years after diagnosis
1986 – “complete remission 6 years and unlikely to relapse”
Quality Improvement

- Collaborative improvement (scientists, paediatricians and haematologists)
- Clinical trials
- Pooling data outcomes
- National level research
Quality Improvements

- Eagles or Weasels
- Jump and grow your wings
- Hunker down or stay safe

Don Berwick 1998
Quality improvements gone wrong

• Eagles or weasels

• Weasels do not fly into the engines of jet aeroplanes

• However weasels spend a lot of time looking down holes in the ground
Paediatric Oncology

Eagles
Child Cancer 0-14 by site

- Leukaemia, 40%
- Brain & CNS, 27%
- Soft Tissue, 9%
- Non-Hodgkin, 9%
- Kidney and renal pelvis, 7%
- Bone & Joint, 6%
- Hodgkin Lymphoma, 4%
- Bone & Joint, 6%
- Kidney and renal pelvis, 7%
- Non-Hodgkin, 9%
- Soft Tissue, 9%
- Brain & CNS, 27%
New Zealand

• 180 diagnoses per annum
• 2 centre model Starship and Christchurch
• 60/120 split
• Collaboration between centres – Parent Information currently working on treatment guidelines
• Thus a National Service model.
Patterns of childhood cancer

• Most malignancies in adults are carcinomas (epithelial tissue)
• More embryonal in children (fetal cells)
• This disparity is usual to age 15yrs then epithelial and non-epithelial similar in incidence (15-19yrs)
• After 19yrs epithelial cancers predominate
Diagnostic issues

- Arise from deep seated tissue therefore not obvious until they are very large.
- No screening techniques - impractical
- Thus approx 80% children have metastases or systemic disease at time of diagnosis
- Multimodal therapy with a MDT approach
Risk Factors

• Unknown
• Genetic factors and certain pre and post natal exposure can increase risk
• No single study can prove causative factors only show risk association but adds to body of knowledge
• Studies show not all children develop cancer for the same reason it is a multi-factorial etiology
Chromosomes
Autosomal dominant cancer syndromes

- Familial retinoblastoma: RB1
- Li-Fraumeni: TP53
- Wilms tumour: WT1
- Breast & ovarian: BRCA1 & 2
Acute Lymphoblastic Leukaemia

- 25% of all childhood cancers
- Sharp peak in children 2-3yrs more boys 2:1
- Mathematical model indicates a “two hit” theory
- Higher in developed world
- 80% survival overall increase due to CNS prophylaxis and treatment intensification.
Survival

• Strongly related to age at diagnosis
  • 1-4yrs – 85%
  • 5-9yrs – 80%
  • 10-14yrs – 68%
  • 15-19yrs – 51%
• Infants have poorest survival at 37%
• Very dependent on biologic sub types
Types

- B Cell – bone marrow
- T Cell – thymus
- Spontaneous mutation in lymphoid cells of B or T cell lineage during normal lymphoid cell development.
Presentation

• Often symptomatic for a number of weeks before diagnosis
• Fever, fatigue
• Bone pain
• Bleeding, petechiae, purpura, pallor
• Lymphadenopathy
• Hepatosplenomegaly
• Infection
Diagnostic tests

- History & Physical
- CBC with diff/Biochemistry
- BMA/LP (CNS involvement)
- CXR
- Renal USS
- Insertion of portacath
The Diagnosis

• The most difficult time of the cancer journey
• Initial symptoms mimic benign problems eg infection/injury
• Often a delay in diagnosis
• S & S related to child and age and tumour type
Clinical Trials

• Family in crisis – informed consent
• Understanding randomisation
• Altruistic intent

• Despite this MOST families commit it is the minority that don’t
• Children’s Oncology Group
• Incremental changes not many new drugs often just a change in delivery.
Treatment plan for ALL

- 2-3 years – longest of all treatments
- Induction (steroids, VCR, +/- Dauno & Lasp)
- Consolidation – CNS prophylaxis ITMtx
- Interim Maintenance – HDMtx/Capizzi
- Delayed Intensification
- Maintenance
Solid Tumours

• Used to do surgery first
• Follow up with chemotherapy – not knowing whether microscopic cells would respond to chemotherapy
• Now, chemo first approx 10 weeks then surgery estimate % tumour necrosis if 90% continue same regimen of drugs
• If poor response then opportunity to change.
Psychosocial impact on the child

• Focus for nurses – identify periods of distress and mediate these issues
• Promote child’s mental health and development
• Shock disbelief and inconvenience
• Constant fear of life threatening illness
• Peak times are diagnosis, induction, completion of therapy
• Relapse
• Changes in body image
• Side effects
• Perceived vulnerability (regression)
• Disruption of usual routines/separation anxiety and abandonment
• Developmental issues
• Incongruence amongst parents
• “Watershed” – end of treatment
• Positive aspects
• Relapse
Psychosocial issues

• Siblings
• Parents
• Grandparents
• Peers/friends
• Financial
• Separation from neighbourhood and supports
Late Effects Assessment Programme (LEAP)

• With overall survival at 80% we know that there will be approx 1:650 youngsters who will be survivors of cancer.

• We also know that with various polymorphisms some children will be affected more by their treatment than others.
Toxicities

- Endocrine
- Fertility
- Sensory
- Organ toxicity
- Neuropsychological
- Mobility
- Cosmetic
- Social

- 58% will have 1 major chronic problem
- 32% will have 2 or more major chronic problems
20 years on from Tx completion

• 90% survival for females
• 87% survival for males
• This is the same incidence as the original cancer!
New agents

- Haematopoietic growth factors
- Peripheral stem cell transplantation
- Interleukins
- Monoclonal antibodies
- Interferons
- Retinoids
- Genotyping
2020

- Changing model of service delivery
- Continuity of care
- Holistic, family centred care
- Nursing leadership
- Development of NP role in CHOC
- Primary care involvement/partnership
Children’s nursing is…….