Adult Childhood Survivorship Program

Provincial Survivorship and Primary Care Program

Survivorship Forum
June 8, 2016
Presenter’s disclosure

• Relationships with commercial interest
  – None
Welcome!

• Matt Hallat
  – Spokesperson
LEAF Team

• Karen Goddard MD
  – Medical Director
• Avril Ullett
  – Program Leader
• Beverley Biggs
  – Counsellor
• Kimberley-Anne Reid
  – Nurse Practitioner
Overview

• Late Effects of Childhood Cancer and Treatment

• Adult Childhood Cancer Survivors Program
  – Program Development
  – Program Components

• Late Effects, Assessment and Follow-up (LEAF) Clinic
Survival Rates

[Diagram showing childhood cancer survival rates over years, with different colors representing various types of cancer such as M. Hodgkin, Wilms Tumour, Acute lymphoblastic Leukemia, Non-Hodgkin Lymphoma, Ewing Sarcoma, Osteosarcoma, Rhabdomyosarcoma, Malignant Germ Cell Tumours, Neuroblastoma, Brain Tumours, and Acute myeloid Leukemia.]
Improvement related to:

• Multimodality approach:
  – Surgery
  – Systemic therapy (chemotherapy)
  – Radiation therapy

• Therapy intensification
  – Bone marrow transplant
  – Interval compression of chemotherapy

• Better supportive care during therapy

• Development of new targeted therapeutic agents
Childhood Cancer is a rare disease

New Cancer Diagnoses in BC in 2009
- Adults 18,815
- Children 145

Why is it so important what happens to these patients?
Incidence

- About 12,000 children in the US (birth – 14 years)
- In Canada 1,310 patients (0-19 years)
- Over 83% of these patients are cured of their cancer
- In 2010, 1 in 250 of the adult population in North America was a survivor of childhood cancer
- Over 3000 survivors of childhood cancer in BC since 1980. These patients are now aged 1-50.
- Life years saved by treating childhood cancer – 2nd only to breast cancer
Survivorship

In memoriam
Ellen Stovall
1946-2016
Late Effects

**Definition:** side effects that occur within more than 5 years after diagnosis.

- At age 45 years:
  - 95.5% cumulative prevalence of any chronic health condition
  - 80.5% for a serious/disabling or life-threatening chronic condition
Late Effects

- Late effects include:
  - Physical problems
    - Organ damage
      - Development affected
      - High risk of late effects in adults treated for childhood cancer
  - Secondary tumors
  - Psychological problems
    - Depression, anxiety
    - Career and financial
    - Relationships
Survivors at Risk

Researchers followed more than 1,700 adults who had been treated for cancer as children and found that those who had received certain types of treatment were very likely to develop certain health problems later in life.

**Adult condition: Breast cancer**

**Childhood treatment:** Radiation to the breast (females only)

**Heart-valve disorder**

Radiation to the heart

**Pituitary dysfunction**

Radiation to the hypothalamus-pituitary

**Hearing loss**

Radiation to the ear or exposure to cisplatin or carboplatin

Sources: St. Jude Children’s Research Hospital; JAMA

The Wall Street Journal
Tumor Related Damage

Invasion into and pressure on different structures

- **Wilms tumor**
  - One kidney usually completely destroyed by disease and has to be removed
Tumor Related Damage

- **Craniopharyngioma** tumor growth and cyst expansion leads to compression of:
  - Optic apparatus
    - Blindness
  - Pituitary
    - Endocrinopathy
Surgery Related Damage

• Surgery
  • Prime modality for local control
• Lymph node dissection
  • Lymphedema
• **Splenectomy**
  • Life threatening infection
    Pneumococcal vaccine
    Medic Alert bracelet
Chemotherapy Related Damage

- Chemotherapy prime modality for systemic control
- Depends on agent and sensitivity of target organs
  - Adriamycin: cardiomyopathy
  - Cisplatin: nephrotoxicity and hearing loss
  - Alkylating agents: infertility and second cancers
  - Vincristine: peripheral neuropathy
Radiation Therapy (RT)

- In children (unlike adults) affects normal growth/development
- Severity of late effects depends on:
  - Age at the time of therapy
  - Total dose given
  - Fractionation
  - Region treated:
    - Some organs more sensitive and easily damaged
    - Amount of normal tissue treated
  - Concurrent chemotherapy can sensitize normal tissues
  - Underlying genetic problems:
    - Radio-genomics
Determining the Risk

• Adult survivors of childhood cancer may be at:
  • Low risk
    – Many patients treated for leukemia
    – No radiation therapy
    – Combination low dose chemotherapy
  • Intermediate risk
    – More intensive chemotherapy
  • High risk
    – Intensive chemotherapy (including alkylating agents and adriamycin)
    – Any previous history of radiation therapy
Organs at Risk

- Central nervous system
- Orbit
- Hearing
- Peripheral Nervous system
- Endocrine
- GU system
- Respiratory
- Gastro-intestinal
- Musculoskeletal
- Reproductive organs
- Cardiovascular
- Skin
Musculoskeletal

• Bone/Muscle/soft tissues
  – “Hypoplasia” – reduced growth within the RT field
Endocrinopathy

- Pituitary dysfunction
  - GH
  - TSH
  - FSH & LH
  - ACTH
Reproductive system

- Gonads very sensitive to both RT and chemotherapy
  - Alkylating agents
  - RT to ovaries:
    - The dose of RT needed to destroy 50% of the oocytes = LD50
    - Oocytes are very sensitive with an LD50 of < 200 cGy
  - Damage to developing uterus
Metabolic Syndrome

- Associated with treatment for childhood cancer
- Cranial radiation therapy and TBI (whole body RT prior to transplant) significantly increase the risk

Etiology
- Poorly understood post chemotherapy alone
- Radiation therapy:
  - Hypothalamic effect
  - Radiation therapy to pancreas

Characterized by:
- Central obesity
- Hypertension
- Hyperlipidemia
- Diabetes
Other CNS Problems

- Brain
  - Developmental delay
    - Poor short term memory
    - Poor executive function
  - Seizures
  - Cerebrovascular events
    - Vascular malformations
    - Early aging of small blood vessels
    - Thrombotic and haemorrhagic
Hearing Loss

- Radiation Therapy:
  - Conductive: wax build up
  - Sensorineural: direct damage to cochlea
- Chemotherapy:
  - Sensorineural
  - Cisplatin causes high frequency hearing loss

Sensory hair cells in the cochlea
Visual Problems

- Cataracts
Dental Problems

• Radiation therapy is associated with:
  – Developmental abnormalities involving roots and enamel (Rx at young age)
  – Xerostomia
    • Due to parotid gland damage
    • Associated with dental decay
  – Trismus
  – Damage to the blood supply of the mandible

• Chemotherapy
  – Developmental abnormalities involving roots and enamel (Rx at young age)

Dental extraction after high dose RT to the jaw can be associated with osteoradionecrosis
Second Neoplasm

- A second cancer or second malignant neoplasm (SMN) is defined as a histologically distinct second cancer that develops after the first.

- **Definition:**
  - Tumor in new location and not from direct spread or metastasis of the primary cancer
  - Tumor in the same location as the primary cancer but of different histological type
Incidence

- Childhood Cancer Survivor Study
  - 30 year cumulative incidence of second malignancy = 9%
Thyroid cancer
Thyroid Cancer Screening

- Check routine thyroid function tests (serum TSH & T4) at least every year
- Examine thyroid every year for nodules
- Thyroid ultrasound scan every 3 years
  - Shows thyroid parenchyma in more detail
  - Suspicious nodules can either be followed closely to look for growth or ultrasound guided fine needle aspiration (FNA) biopsy can be organized
Radiation-induced Meningioma

- **RT induced meningioma**
  - Multiple
  - Atypical
  - More likely to recur after surgery
Screening for Meningioma

• MR scan every 3 – 5 years
Skin Cancer

- Increased risk of cancers in previous radiation therapy field
  - Basal cell carcinoma
  - Melanoma
Skin Cancer

- Screening:
  - Careful examination of skin in previous RT field
  - Previous history of chemotherapy: Look for melanocytic lesions
- Prevention:
  - Hat, T-shirt, sunscreen
  - “Dear 16 year old me”
Colorectal Cancer (CRC)

- 2-3% risk of CRC 30 – 40 years after treatment for childhood cancer and increasing.
- Associated with abdominal radiation therapy
- Risk is proportional to dose and volume of RT
  - Increased by 70% with each 10-Gy increase in RT dose.
  - Increased RT volume increased risk (group 1 OR, 1.5; P .001; group 2 OR, 1.8; P .001).
- Alkylating agent exposure associated with 8.8X increased risk of secondary CRC.
Secondary Breast Cancer

- Commonest solid tumor among female survivors of Hodgkin lymphoma
- Moderately high-dose mediastinal RT
  - Scatter to adjacent (breast) tissue
- Adolescent girls most at risk
Secondary Breast Cancer

- After treatment for Hodgkin lymphoma in adolescence
  - 37X risk of breast cancer
  - Bilateral disease more common
  - Increased risk:
    - Over 12 years of age at diagnosis
    - Higher dose of radiation

Unilateral and bilateral breast cancer in women surviving pediatric Hodgkin's disease.
Breast Cancer Screening

• Who is this recommended for?
• Women who received:
  – RT associated with significant scattered radiation to the breast
  – Doses of 20 Gy or higher to the following fields:
    • Mantle
    • Mediastinal
    • Whole lung
    • Axillary fields
Breast Cancer Screening

- Recommendations:

<table>
<thead>
<tr>
<th>Problem</th>
<th>Screening/Investigation</th>
<th>Frequency</th>
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</thead>
<tbody>
<tr>
<td>Increased Breast cancer risk</td>
<td>Breast self examination</td>
<td>monthly</td>
</tr>
<tr>
<td></td>
<td>Breast examination by HCP</td>
<td>Annually until aged 25 and then 6 monthly thereafter</td>
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<td></td>
<td>Breast MR</td>
<td>Annually starting at age 25 or 8 years after the RT was given</td>
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<tr>
<td></td>
<td>Mammograms</td>
<td>Annually starting at age 25 or 8 years after the RT was given</td>
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Screening

• Screening guidelines available
  – Children’s Oncology Group

Health Link

Thyroid Problems after Childhood Cancer

Some people who were treated for cancer during childhood may develop endocrine (hormone) problems as a result of changes in the function of a complex system of glands known as the endocrine system.

What is the endocrine system?

The endocrine system is a group of glands that regulate many body functions including growth, puberty, energy level, urine production, and stress response. Glands of the endocrine system include the pituitary, hypothalamus, thyroid, adrenals, pancreas, ovaries (in females), and testes (in males). The hypothal-
Radiation-induced GBM
Screening

• Screening not possible for all late effects:
  – Some radiation induced malignancies such as sarcomas and malignant brain tumors
Psychological Risk Factors:

• Increased psychological distress: depression, anxiety and PTSD

• Decreased health-related quality of life

• Interpersonal relationship issues

• Slower cognitive processing
Many brain tumor survivors:

• Need very modified school programs and support
• Rely on permanent disability pension:
  • Differences across the province and between different provinces regarding available programs
    • Access to vocational/recreational rehab
• Drug costs covered by parents benefits plan
• Other costs not covered:
  • Hearing aids
Many Survivors of Childhood Cancer:

• Engage in Risky Health Behaviours:
  • Smoking
  • Alcohol and Drug use
  • Altered sleep patterns
  • Suffer from Chronic Fatigue
Other Psychosocial Issues:

• Low educational attainment

• Low household income

• Unemployment

• Unmarried/partnered

• Less likely to achieve independent living

• Less likely to be in a long term relationship
Other Issues:

- Fertility
- Sexuality
- Body image

- Poverty and affordable housing
Impact on Life

• Huge range of late effects:
  – Low risk:
    • Many (but not all) previous lymphoma and leukemia patients
    • Function very well
    • Minimal risk for long-term health problems
  – High risk:
    • Any radiation, high dose chemotherapy - including alkylating agents and anthracylines
    • Some leukemia patients, brain tumors and solid tumors (e.g. sarcomas)
    • Lives may be “devastated”
  – Long term health care:
    • Counseling
    • Screening/Surveillance for late effects
Screening

- Generally, follow up care depends on “risk category”
  - High risk: Hospital based and family practitioner
  - Low risk: Family practitioner
- Survivorship Care Plan:
  - Coordinated post-treatment plan
  - Built by survivor’s oncology team
  - Includes
    - Summary of the survivor’s treatment
    - Direction for future care
- Screening recommendations: COG Long Term FU Guidelines
History

- History of previous illness and therapy critical to understanding risks and organizing appropriate screening
- Generate a “care plan”
Prevention

Information about late effects critical for prevention:

• Initial therapy
  – Give treatments which are less likely to cause long-term damage
    • Avoid or reduce radiation therapy
    • Targeted therapy
      – We don’t know about the late effects of these agents yet
  – Tailored therapy
    • Genomic studies to identify people more likely to develop side effects
Prevention

- Information/education
  - Childhood cancer survivors
    - Know how to seek advice
  - Health care Professionals
    - Do the correct investigations
Prevention

• Lifestyle:
  – Diet
  – Exercise
  – Smoking
  – Sun/UV exposure
Cancer Survivors Who Stay Active Live Longer

By CRETCHEN REYNOLDS  MAY 16, 2012 12:01 AM  81 Comments
Prevention is Complex

- **Risk of cerebrovascular disease:**
  - Radiation therapy (RT)
    - Dose, fractionation and area treated
  - Chemotherapy at time of RT
  - Genetic factors
  - Endocrinopathy
    - GH deficiency
  - Metabolic syndrome
    - Hyperlipidemia
    - Hypertension
    - Diabetes
  - Lifestyle
    - Smoking and exercise
  - Genetic factors
  - Prophylactic aspirin
    - Hemorrhagic and thrombotic
- **Publication just accepted:**
  - A cross-sectional cohort study of cerebrovascular disease and late effects after radiation therapy for craniopharyngioma
Resources

COG: Long-Term Follow-Up Guidelines for Survivors of Childhood, Adolescent, and Young Adult Cancers
Resources

– National Cancer Institute:

Late Effects of Treatment for Childhood Cancer
Resources

Pediatric Oncology Education Materials

Late Effects

General Overview

On average approximately 10,400 North American children (between birth and 14 years of age) develop childhood cancer each year and these numbers seemingly increase annually.1

More than 80% of these children will be long term survivors who have been cured of their disease. This was very different 20 to 30 years ago, when many children did not survive.2

In general, cure rates have been improved by using:

- Multiple treatment modalities
  - Radiation therapy (RT)
  - Chemotherapy
  - Surgery
- Therapy intensification (using higher total doses of chemotherapy over a shorter period of time)3
- Improved supportive care

BC Cancer Agency
CARE + RESEARCH
An agency of the Provincial Health Services Authority
Adult Childhood Cancer Survivors Program
Program Development

January 2014: task group formed
- Survivors and family members of survivors, community family physicians, and representatives from BCCA and BCCH including oncologists, senior leaders, survivorship, researchers, and nursing.

March-October 2014: monthly meetings to develop program

October 2014: business case finalized and submitted to PHSA executive

November 2014: business case sent to Ministry of Health

January 2015: Ministry of Health, BC Cancer Foundation and BC Children’s Hospital Foundation agree to fund program
Program Components

• Multidisciplinary, specialized Late Effects, Assessment and Follow-up (LEAF) clinic

• Recall program

• Transition program

• Registry

• Primary care, patient and family education and support

• Primary care attachment and support
Program Timeline

Start-up (Year 0)

• Recruit and train staff; establish program governance; develop engagement, communication, education, research and evaluation plans
• Define, plan, implement recall process

Year 1

• Recall initiated for high risk patients
• Patients transitioning out of BCCH are stratified by risk and transferred to appropriate provider
• High risk and complex patients seen in Specialty Clinic

Year 2

• Recall for high risk patients completed

Year 3

• Data analysis begins
• Recall for low and moderate risk patients begins

Year 4 and 5

• Recall complete
• Program evaluation report complete
Province ACCS Survivorship Program

Tier One
- Primary Care
  + 25 new low risk patients per year
  + low risk post recall

Tier Two
- Primary Care with Specialized Knowledge and Training
  + 35 new moderate risk patients per year
  + moderate risk post recall

Tier Three
- Specialty Care
  + 50 new high risk patients per year
  + high risk post recall

Tier Four
- Specialty Care
  ACCS with existing side effects
  + 10 new complex high needs new patients
  + no new post recall

Specialized Support

Specialized follow up

Targeted follow up

Generalized follow up

BC Cancer Agency
ACCS (Adult 17+)
Survivorship

BC Children's Hospital
ACCS (Adult 17+)
Transition Services
LEAF Clinic Visit - Medical

• Review of past cancer diagnosis and treatment
  – Review patient summary
  – Patient receives copy of summary
• Assessment of past, current and chronic health problems
• Discuss future health risks
• Make plan for surveillance and treatment
• Order investigations as required
• Summary consult back to primary care provider and involved specialists
LEAF Clinic Visit - Psychosocial

- Consultation and assessment
- Short-term counselling
- Case management
- Advocacy and referral to other BCCA and BCCH programs, and community programs and health providers
LEAF Clinic Psychosocial Support:

- Advocacy with the Ministry of Social Development re: MNS and Non-local medical transportation Requests

- Information and advice on future planning:
  - BC Housing/Supportive Housing
  - RDSPs
  - Representation Agreements
  - Advanced Care Planning.
LEAF Clinic Psychosocial Support:

• Support provided province-wide with a variety of modalities
  – In person
  – Telephone (telephone counselling to come at a later time)
  – Email resources and link to appropriate resources
Eligibility

- Diagnosed age 17 and under
- Currently over age 18
- Currently 5 years off of active treatment
- Already discharged from BC Children’s Hospital
Referrals to LEAF Clinic

- BC Children’s Hospital
- BC Cancer Agency
- Primary Care Providers
- Allied Health/Community Referrals
- Self-referrals
Thank you!

Any questions about the care of an adult childhood cancer survivor under 21 years of age?
• Contact:
  – Sheila Pritchard
    • spritchard@cw.bc.ca
  – Marion Nelson (nurse practitioner)
    • mnelson@cw.bc.ca

Any questions about the care of an adult childhood cancer survivor over 21 years of age?
• Contact:
  – Karen Goddard (radiation oncologist)
    • kgoddard@bccancer.bc.ca
  – Kimberley-Anne Reid (nurse practitioner)
    • Kimberley-Anne.Reid@bccancer.bc.ca
  – Beverley Biggs (Social Worker)
    • Beverley.Biggs@bccancer.bc.ca
  – Avril Ullett (Program Leader)
    • Avril.Ullett@bccancer.bc.ca
What didn't kill me made me Stronger