The New Patient:

Work-up and Management

CONTENTS

ASSESSMENT AND INVESTIGATION OF CANCER IN CHILDREN

History

In addition to a detailed history of the presenting complaint and systematic inquiry, make sure you have covered the following:

- Detailed family history particularly of cancers – multiple cancers in individuals (eg. bilateral breast cancer) and cancers occurring in folk < 50 years old are of particular interest
- Ask about DVT and bleeding tendencies in the family
- Detailed history of immunisations and viral exanthema especially chickenpox and measles. Have siblings been immunised?
- Family relationships including names and ages of parents and all siblings. Are there any pre-existing psychosocial problems?
- Ask about pain. Parents may not be aware that their child has been in pain so specifically ask about disrupted sleep, wincing when picked up etc.

Examination

GENERAL:

- Plot weight and height on percentile chart
- Temperature
- Evidence of cachexia
- Pallor, jaundice, oedema
- Phenotypic features eg. features of Beckwith-Weidemann syndrome, isolated hemihypertrophy, Downs syndrome (mosaics with subtle abnormalities may develop leukaemia), café au lait pigmentation etc
- Bruising and petechiae

CARDIOVASCULAR SYSTEM

- Perfusion, BP, pulse, auscultation
- Evidence of SVC obstruction for patients with anterior mediastinal masses (dilated neck veins, swollen eyelids/face, dusky face)
- Evidence of pericardial effusion with chest masses

RESPIRATORY SYSTEM

- Evidence of distress, cyanosis
- ? tracheal obstruction/deviation with anterior mediastinal masses
- Evidence of pleural effusion with chest or abdominal masses

ABDOMEN

- General examination – measure enlargement of liver, spleen, masses
- Bimanual palpation for renal masses

NEUROLOGICAL

- General examination
- Fundoscopy and careful examination of 7th and 3rd cranial nerves for patients with leukaemia
- Careful assessment of bulbar function for patients with brain stem tumours or cancers eroding through the base of the skull
- ? Horner's syndrome with neck and posterior mediastinal tumours (smaller pupil, apparent enophthalmos, but heterochromia unusual as this usually reflects a prenatal origin)
- assessment of limb strength, sensory level, bowel and bladder in patients with spinal/paraspinal masses

GENITO-URINARY:

- Evidence of precocious puberty (germ cell tumour, hepatoblastoma)
- Testicular enlargement (germ cell tumour, ALL)
- Undescended testes (dysgerminoma)
- Other developmental abnormalities – Wilms

LYMPH NODES

- Feel carefully for all groups including cervical, pre/post auricular, supraclavicular, axillary, inguinal and around knee and elbow joints
- It is surprising how often obvious supraclavicular glands are missed – there is a fullness above the clavicle but you cannot feel the lower border of the gland
- Left supraclavicular gland (Virchow Trosier) in patients with abdominal masses (particularly neuroblastoma)
- Sometimes massive cervical adenopathy is missed because the gland is deep to the sternocleidomastoid muscle – it may be easier to see it!
- Remember that Waldeyer's ring is part of the lymphoid system – examine carefully in patients with neck masses

ENT

- Examine for dental problems
- Gum hypertrophy and infiltration may occur with AML, JMML, LCH
- Evidence of middle ear disease which could signal problems when neutropenic.
- Sarcoma can arise from the middle ear

EYES

- Aniridia with Wilms
- Fundal/subconjunctival haemorrhage with leukaemia

Routine investigations, consultations and supportive care on all patients

HAEMATOLOGY

Full blood count, differential and smear

COAGULATION

PT, APTT, thrombin clotting time and fibrinogen

D-dimers only if suspicion of DIC

"thrombophilia screen" if history strong for DVT

BLOOD BANK

group and save

BIOCHEMISTRY

Na, K, urea, creatinine, glucose, bilirubin, ALP, GGT, AST, ALT, Ca, Mg. PO_4, albumin, urate, LDH

DENTAL ASSESSMENT

HEARING

MOUTH CARE

commence appropriate care when diagnosis established (see Symptom Care chapter)

SUPPRESSION OF MENSTRUATION

See section in Symptom Care chapter.

UNIT CHILD CANCER REGISTRY

All families are requested to consent for data on their child's cancer to be submitted to the NZCCR.

Suspected leukaemia

In addition to the above, do:

- Blood
- Chest X-ray
  - PA + lateral (? mediastinal/hilar mass)

- Ultrasound abdomen
  - ? renal infiltration
- ? febrile
  - investigate and treat as for febrile neutropenic patient regardless of the neutrophil count.
- BM aspirate

  Presentation marrows may be very difficult to aspirate
- BM trephine
  - Only obtain if aspirate of poor quality
  - Roll on slide and rest in formalin
- LP
  - Ensure platelet count > 50
  - Collect 20 drops for cell count and cytospin
  - Only instil intrathecal methotrexate at time of initial LP if certain that diagnosis is one of ALL from blood cell markers and microscopy

- Hyperhydration - see Tumour Lysis Syndrome in Emergencies chapter

  And allopurinol
- Packed cell transfusion
  - Keep Hb > 80 but not absolutely necessary before BM aspirate and beware transfusion of packed cells if presenting white cell count > 100

Suspected lymphoma

In addition to the routine investigations done on all patients. Depends on type suspected:

T-cell NHL

- as for suspected leukaemia above but
- beware superior mediastinal syndrome (see Emergencies chapter)
- requires lymph node or mediastinal mass biopsy

B-cell NHL

- as for suspected leukaemia above
- CT chest, abdomen and pelvis
- be particularly aware of Tumour Lysis Syndrome (Emergencies chapter)

- avoid laparotomy; diagnosis can often easily be established by tapping pleural effusion or ascites
- CT/MRI brain if CSF positive

Hodgkin's disease

- CXR (PA + lateral)
- excision biopsy of lymph node
- CT chest and abdomen (ensure good oral contrast)
- BMA + trephine if stage III/IV
- Bone scan is symptomatic
- LP and MRI CNS for paraspinal tumours

Suspected solid tumour

In addition to routine investigations:

- Check Ca^{++} - one is occasionally surprised by hypercalcaemia
- Scan of primary site - CT, MRI or ultrasound
- Chest X-ray (PA + lateral)

Neuroblastoma

INVESTIGATIONS TO CONFIRM DIAGNOSIS AND STAGE

1. spot urine for urinary catecholamines
    - HMMA/creatinine ratio
    - HVA/creatinine ratio
    - Dopamine/creatinine ratio
2. bone scan
3. bilateral posterior iliac crest aspirates and trephines
4. serum ferritin

Wilms tumour

INVESTIGATIONS TO CONFIRM DIAGNOSIS AND STAGE

1. check IVC and contralateral kidney carefully
2. CT chest

Soft tissue sarcoma

INVESTIGATIONS TO CONFIRM DIAGNOSIS AND STAGE

1. bilateral posterior iliac crest aspirates and trephines
2. bone scan
3. CT chest
4. local examination if appropriate eg. cystoscopy/vaginoscopy/ENT

Ewing's sarcoma

INVESTIGATIONS TO CONFIRM DIAGNOSIS AND STAGE

1. bilateral posterior iliac crest aspirates and trephines
2. bone scan
3. CT chest

Osteosarcoma

INVESTIGATIONS TO CONFIRM DIAGNOSIS AND STAGE

1. bone scan
2. CT chest

Hepatoblastoma

INVESTIGATIONS TO CONFIRM DIAGNOSIS AND STAGE

1. CT chest
2. α-fetoprotein and β-HCG
3. Hepatitis serology

Germ cell tumour (extracranial)

1. CT chest
2. α-fetoprotein
3. β-HCG
4. bone scan

Retinoblastoma

INVESTIGATIONS TO CONFIRM DIAGNOSIS AND STAGE

Hereditary	Sporadic unilateral
Ultrasound eyes	Ultrasound eyes
MRI brain (? Trilateral disease)	

Then decide if further investigations are necessary if high risk features present (applies to both hereditary and sporadic cases):

High risk features

(any 1 of following)

1. cut end of optic nerve involved, or
2. involvement of optic nerve beyond the lamina cribrosa, or
3. “massive” choroidal infiltration, or
4. infiltration of sclera, or
5. infiltration into the orbit
6. involvement of the ciliary body or structures more anterior
7. CNS disease on MRI
8. Suspicion of distant spread clinically – enlarged pre-auricular gland, bony pains, depression of blood count

Investigations to be done on all cases

- ❑ MRI brain and spine
- ❑ LP for cytology (Anatomical Pathology)
- ❑ Bone scan
- ❑ Bilateral posterior iliac crest aspirate and trephine biopsy

Brain tumour

ON ALL PATIENTS

- ❑ MRI brain
- ❑ MRI spine if brain scan not suggestive of low grade glioma
- ❑ Discuss with neurosurgeon
- ❑ Commence dexamethasone 0.25 – 1 mg/kg/day in 4 divided doses IV/PO

Wilms Tumour Predisposition Syndromes

The following syndromes/conditions are associated with an increased risk of Wilms tumour and sometimes other types of malignancies:

Syndrome	Risk of malignancy	Other tumours
Beckwith-Wiedemann	7.5%	hepatoblastoma adrenocortical Ca neuroblastoma
BWS with hemihypertrophy	40%	as for BWS
Hemihypertrophy (isolated)	3 - 5%	as for BWS
Sporadic aniridia (ie. non-inherited form) may occasionally be associated with hemihypertrophy	20 - 40%	nii
Denys-Drash syndrome (nephrotic syndrome, male pseudohermaphroditism)	30 - 90%	germ cell tumour (in intra-abdominal dysgenetic gonads)
Perlman's syndrome (gigantism, abnormal facies, renal hamartomas)	? (high)	nil
WAGR syndrome Wilm's, aniridia, Genitourinary abnormalities, mental retardation)	30 - 40%	nil

Wilms tumour has a doubling time of around 1 week. Metastatic Wilms tumour has been diagnosed in-between 6 monthly screening ultrasounds. Greater than 90% of Wilms tumour are diagnosed by the end of the 7th year of life in children with the above predisposition syndromes.

CALCULATION OF BODY SURFACE AREA

Sharkey et al, British Journal of Cancer 85(1): 23-28, 2001

Surface area should be estimated using the table below rounding off to the nearest kilogram ie. 12.5 to 13.4kg is estimated at 13 kg.

Exclusions

- Weight < 10kgs. Most protocols provide specific recommendations for infants.

BSA estimation in patients greater than 10kg

$$(m^2) = \sqrt{\frac{Ht\ (cm) \times Wt\ (kg)}{3600}}$$

BODY WEIGHT (KG)	SURFACE AREA (m^2)
11	0.53
12	0.56
13	0.59
14	0.62
15	0.65
16	0.68
17	0.71
18	0.74
19	0.77
20	0.79
BODY WEIGHT (KG)	**SURFACE AREA (m^2)**
21	0.82
22	0.85
23	0.87
24	0.90
25	0.92
26	0.95

BODY WEIGHT (kg)	SURFACE AREA (m^2)
51	1.5
52	1.5
53	1.5
54	1.6
55	1.6
56	1.6
57	1.6
58	1.6
59	1.7
60	1.7
BODY WEIGHT (kg)	**SURFACE AREA (m^2)**
61	1.7
62	1.7
63	1.7
64	1.7
65	1.8
66	1.8

27	0.97
28	1.0
29	1.0
30	1.1
31	1.1
32	1.1
33	1.1
34	1.1
35	1.2
36	1.2
37	1.2
38	1.2
39	1.3
40	1.3
41	1.3
42	1.3
43	1.3
44	1.4
45	1.4
46	1.4
47	1.4
48	1.4
49	1.5
50	1.5

67	1.8
68	1.8
69	1.8
70	1.9
71	1.9
72	1.9
73	1.9
74	1.9
75	1.9
76	2.0
77	2.0
78	2.0
79	2.0
80	2.0
81	2.0
82	2.1
83	2.1
84	2.1
85	2.1
86	2.1
87	2.1
88	2.2
89	2.2
90	2.2