

# Lymphoma

## Definition:

A cancer of the lymph glands

## **Hodgkin's Disease (HD)**

## Definition:

A cancer characterized by progressive, often painless, enlargement of lymph nodes

## Signs and Symptoms

- Painless lymphadenopathy (cervical lymphadenopathy is most common)
  - ~ Splenomegal
- Systemic or "B" symptoms
  - ~ unexplained fever
  - ~ night sweats
  - ~ weight loss of 10% or more or failure to gain weight in young children

## Diagnostic Evaluation

- CBC, ESR, coagulation profile
- Liver function tests, LDH, renal function
- Serum copper and ferritin (often elevated)
- Bone marrow aspirate and biopsy
- Diagnostic surgical lymph node biopsy
- Chest x-ray
- Abdominal ultrasound
- CT scan of chest, abdomen, +/- neck
- Gallium scan

## Staging

Stage I - 1 lymph node region positive

Stage II - 2 or more lymph node regions on the same side of the diaphragm

Stage III - lymph node regions on both sides of the diaphragm

Stage IV - involvement of extra lymphatic organs

Patients are also classified into "A" - no systemic symptoms, and "B" - systemic symptoms.

## Histology

- Lymphocyte predominant
- Nodular sclerosing
- Mixed cellularity
- Lymphocyte poor

## Treatment

### 1. Acute Management

Airway obstruction or superior vena caval syndrome

- May need intubation or tracheostomy
- May need emergency radiotherapy

## 2. Specific Therapy

Treatment depends on staging.

### a. Radiotherapy

May be used alone in Stage IA and IIA. Radiotherapy should be avoided in very young children because of its effect on growth and chemotherapy should be used instead.

### b. Chemotherapy

May be used for all stages of disease

Always used in advanced stage IIB, III and IV

Most commonly used chemotherapy combinations:

ABVD - doxorubicin, bleomycin, vinblastine, dacarbazine

COPP - cyclophosphamide, vincristine, prednisone, procarbazine

MOPP/ABV "Vancouver Hybrid"

## Prognosis

- Factors associated with a good prognosis:
  - ~ Young age
  - ~ female sex
  - ~ absence of systemic symptoms
  - ~ Stage I or II disease
  - ~ Lymphocytic predominant subtype
  
- Factors associated with poor prognosis:
  - ~ Large mediastinal mass
  - ~ Stage IV disease
  - ~ Lymphocyte poor histology
  
- Overall survival approximately 75%

# Lymphoma

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A cancer of the lymph glands

## **Non Hodgkin's Lymphoma (NHL)**

## Definition:

This is due to a malignant proliferation of cells of the lymphocytic or histiocytic lineage.

## Classification of NHL

Sub Type	Cell Type	Usual Site	Treatment
Lymphoblastic Lymphoma	Immature T-cell (occasionally Pre-B)	Anterior mediastinum	As for T-cell ALL
Small Non-Cleaved	B-cell (Burkitts)	Abdomen - 90%	"LMB-90"
Large Cell (histiocytic)	Immunoblastic B, T Large cell-B Anaplastic Large cell (Ki-1 positive) usually T	Abdomen, mediastinum, skin, CNS, lymph node, testes, bowel	

## Signs and Symptoms

- **Persistent** Lymph node enlargement (the commonest cause of enlarged lymph glands in children is infection)
- Shortness of breath
- Abdominal pain, bowel obstruction
- Occasional fever without signs of infection and night sweats (more common with Hodgkin's Disease)

## Diagnostic Evaluation – at BCCH

- CBC and differential, liver function tests (including LDH), renal function (BUN, creatinine, uric acid, electrolytes)
- Bone marrow aspirate and biopsy. Cytogenetics and immunophenotyping essential. If bone marrow contains >25% lymphoma cells, it is classified as leukemia
- Surgical biopsy of lymph node. Cytochemical, immunologic, cytogenetic and molecular studies essential
- Spinal tap – examine fluid for presence of abnormal cells and molecular studies
- Chest x-ray
- CT scan of abdomen +/- chest
- Gallium scan
- Bone scan

## Treatment

### 1 Acute and Supportive Care

Children may present acutely with severe difficulty breathing. Emergency oncological consultation may be required.

Bowel obstruction or bleeding - Surgical resection if necessary. Resection should preserve as much bowel as possible. It is not necessary to obtain a complete tumour resection

## 2 Specific Treatment.

### a) Chemotherapy

Is the mainstay of treatment. There are many different treatment regimens and the type used depends on the type of NHL and whether or not the disease is localized (most are systemic or advanced at diagnosis). Most commonly used chemotherapy agents for NHL are cyclophosphamide, vincristine, methotrexate, prednisone, doxorubicin

### b) Intrathecal chemotherapy

Patients with non-localized disease are at risk for CNS involvement and therefore intrathecal chemotherapy (via a spinal tap) is used prophylactically

### c) Radiation therapy

1 Used for emergency treatment in life threatening situations

2 May be used for cranial prophylaxis in high risk patients

Radiation therapy alone is not sufficient treatment for non Hodgkin lymphoma in children

### d) Surgery

Surgical biopsy is usually essential for diagnosis. Localized bowel disease may be completely resected but the patient will still require chemotherapy.

## Staging/Grouping

In children the staging system used is “localized” or “diffuse”. The extent of bone marrow infiltration, central nervous system involvement and LDH levels are of prognostic value.

## Prognosis

This depends on the type and stage but overall approximately 70% event-free survival.