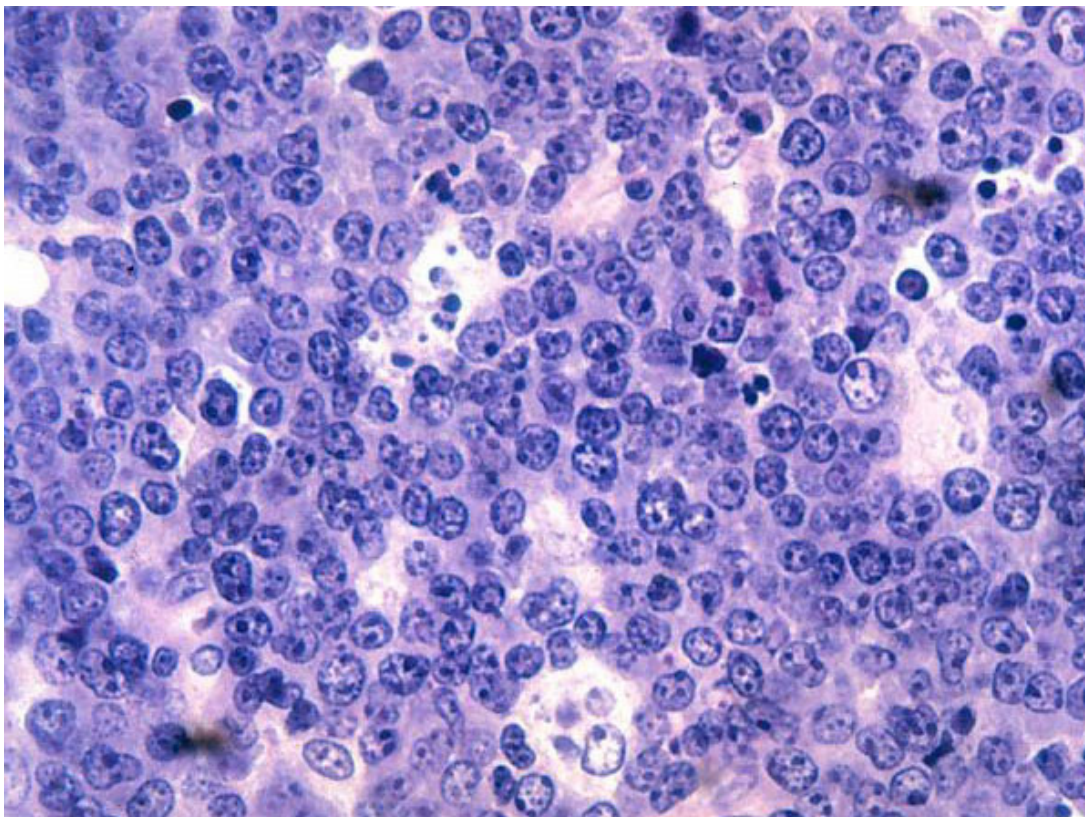


## Burkitt Lymphoma — Causes and Symptoms

[See online here](#)

**Burkitt lymphoma (small non-cleaved cell lymphoma) is one of the most aggressive and rapidly growing non-Hodgkin's lymphomas, with obvious predominance in children and young adults. This type of tumor requires urgent therapeutic intervention as the CNS and the bone marrow are involved in the malignant process since the early stages of the disease.**



### Definition and Background of Burkitt Lymphoma

Burkitt lymphoma (BL) is an aggressive **B-cell** non-Hodgkin lymphoma (NHL). It is named after Dr. Denis Burkitt, who described lymphoma in African children ('African-type' or 'endemic' Burkitt lymphoma). Another type is the 'sporadic' or 'non-endemic' type that is more common in the Western world.

### Epidemiology of Burkitt Lymphoma

The incidence of endemic and sporadic Burkitt lymphoma (BL) varies geographically.

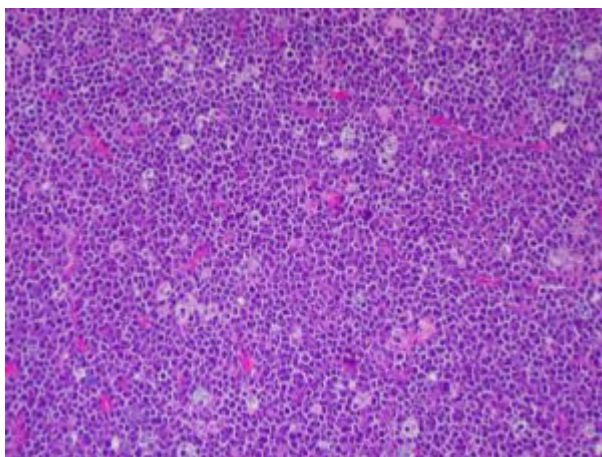
The **endemic BL** is found in Africa, hence also called 'African' Burkitt lymphoma. The highest incidence of BL occurs in children aged 4-7 years, with an estimated incidence of

4-5 cases per 100,000 children annually. BL accounts for one-third to one-half of all childhood cancers in equatorial Africa. Males are twice more commonly affected than females.

The **sporadic** or **non-endemic BL** is common in the United States and Western Europe. It comprises 30% of pediatric lymphomas and less than 1% of adult NHLs in the US, with an estimated incidence of 3 cases per million persons annually. The sporadic BL is also more common in males with an M:F ratio of 3-4:1.

## Etiology and Pathophysiology of Burkitt Lymphoma

The cancerogenic process of BL takes place in the **germinal centers of the lymphoid organs** (Peyer's patches, **spleen** and lymph nodes). In BL, a characteristic genetic mutation is observed, i.e., **translocation** and **deregulation** of the **c-MYC gene** (a proto-oncogene) on chromosome 8, which is responsible for the development of BL.



Burkitt lymphoma, by Ed Uthman from Houston, TX, USA, H&EUploaded by CFCF, CC BY 2.0, <https://commons.wikimedia.org/w/index.php?curid=30103521>

In nearly all cases of endemic (African) BL, **Epstein-Barr virus** infection (the cause of infectious mononucleosis) is found; such a clear association is not found in cases of the sporadic BL.

Histologically, BL manifests as a "starry sky" appearance, with the monoclonal proliferation of medium-sized, non-cleaved B-cells that are similar in shape and size; they make up a diffuse pattern of tissue engagement.

## Clinical Presentation of Burkitt Lymphoma

The Burkitt lymphoma is a very aggressive B-cell lymphoma that presents with rapidly growing cancer masses that have very short doubling times. The constellation of symptoms of BL depends upon the scale of the lesion, CNS involvement and the effect on contiguous organs.



Picture of the mouth of a patient with Burkitt lymphoma showing disruption of teeth and partial obstruction of the airway," by Mike Blyth - Own work. License: CC BY-SA 2.5

The endemic (African) BL commonly presents with a large mass involving the mandible or maxilla. The primary abdominal and bone marrow involvement is less common.

The sporadic BL often presents with primary abdominal involvement, such as abdominal masses, distension, and ascites involving the stomach and intestines. The bone marrow is also involved in about one-third of the cases.

The BL, like other NHLs, is associated with **painless lymphadenopathy**, which is more common in adults rather than in children. Patients with BL may present with the constitutional symptoms, called **B-symptoms**, such as fatigue, night sweats, loss of weight. They are often exhausted, complain about vomiting, thirst, and dizziness. The condition may also manifest like a severe intoxication with signs of **kidney failure**. All symptoms are characterized by swift turnover of mature B-lymphocytes.



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#### The other clinical features of BL are:

- Abdominal masses
- Ascites
- Hepatosplenomegaly
- Ecchymosis and/or petechiae (as a result of thrombocytopenia) "dot-like" skin

hemorrhage

- Meningeal signs (from CNS disease), confusion, headaches, impairment of eyesight and paraplegia

## Laboratory studies

- **Excisional biopsy** (important for confirmation of diagnosis).
- **Complete blood cell (CBC) count** with differentials:
  - In the case of bone marrow involvement, there is **pancytopenia**.
- **Coagulation tests** (prothrombin time [PT], partial thromboplastin time [PTT] and fibrinogen) in order to detect and evaluate the probability of disseminated intravascular coagulopathy (DIC).
- **Serum electrolyte levels:**
  - In BL, there is hyperkalemia, hyperphosphatemia, hyperuricemia, and **hypocalcemia**.
  - If uric acid levels are high, this is evidential of high-grade malignancy.
  - Lactate dehydrogenase is extremely elevated.
  - Creatinine (kidney function tests).
  - The abnormal serum electrolytes, a very high serum lactate dehydrogenase (LDH) levels and hyperuricemia may occur due to **spontaneous tumor lysis**.
- **Liver function tests**
- Tests for **human immunodeficiency virus (HIV)** and **hepatitis B (HBV) infection** for all patients.
- **Beta2 microglobulin** is a sign of extension of the disease and its relapse.
- **Immunophenotype** and **cytogenetic studies**.

## Imaging studies

- Head or spinal computed tomography (CT) scanning;
- Magnetic resonance imaging (MRI);
- Bone scanning and plain bone radiography.

## Staging of Burkitt Lymphoma

### St. Jude/Murphy staging system for Burkitt lymphoma

Stage I	There is a solitary tumor (extranodal) or single anatomic area (nodal); abdomen and mediastinum are not involved in the process.
Stage II	<ul style="list-style-type: none"><li>• Single extranodal tumor with involvement of local nodes.</li><li>• Two extranodal tumors on the same side of the diaphragm, with—or without—regional node involvement.</li><li>• Primary gastrointestinal tumor, with—or without—involvement of mesenteric nodes only.</li><li>• Two or more nodal areas on the same side of the diaphragm.</li></ul>
Stage IIR	Completely resected intra-abdominal disease
Stage III	<ul style="list-style-type: none"><li>• Two single extranodal tumors on opposite sides of the diaphragm;</li><li>• All intrathoracic tumors (mediastinal, thymic and pleural);</li><li>• All paraspinal and epidural tumors, regardless of any other tumor sites;</li><li>• All extensive primary extra-abdominal diseases;</li><li>• Two or more nodal areas on the opposite sides of the diaphragm.</li></ul>
Stage IIIA	Localized but non-resectable intra-abdominal disease.



Stage IIIB	Widespread multiorgan abdominal disease.
Stage IV	Any of the above with initial CNS and/or bone marrow involvement (less than 25% involvement; greater than 25% involvement is defined as L3ALL-acute lymphoblastic leukemia).

## Management of Burkitt Lymphoma

The mainstream therapy in the management of Burkitt lymphoma is **chemotherapy**. The common chemotherapeutic agents used to treat BL include:

- **CODOX-M/IVAC** (Magrath regimen) (cyclophosphamide, vincristine, doxorubicin, high-dose methotrexate / ifosfamide, etoposide, high-dose cytarabine) and the CALGB 9251 protocol-intensive and short-term therapy.
- **Hyper-CVAD** (modified fractionated cyclophosphamide, vincristine, doxorubicin, dexamethasone) and the Cancer and Leukemia Group B (CALGB) 8811 protocol-long-term therapy.
- **Rituximab** is added to mainstream therapy.
- **Urate-oxidase enzymes** (rasburicase).
- **Glucocorticoids** (prednisone as the remedy for immunosuppression).

**Autologous stem cell transplantation** can be performed in the patients during the early stages of the disease with promising results.

Radiotherapy and surgery are not the recommended options in Burkitt lymphoma.

## Supportive Therapy

As the **hemopoiesis** is oppressed in Burkitt lymphoma, the transfusions of the blood elements are compulsory (red blood cells or platelets) in the case of **anemia** and **thrombocytopenia**.

**Intravenous antibiotics** are administered for neutropenic fevers and CNS involvement.

Intravenous fluids are administered to adequately **hydrate** the patient and maintain high urine output in order to prevent **tumor lysis syndrome**.

## Long-term monitoring

Long-term monitoring is compulsory in case of Burkitt lymphoma as any other malignant tumors; the patients have regular checkups with their oncologist.

## References

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Notes