Tikur Anbessa Specialized General Hospital
Pediatric Hematology/Oncology

NURSE RESOURCE BINDER

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SIGNS OF CHILDHOOD CANCER

C ontinued, unexplained weight loss
H eadaches, often with early morning vomiting
I ncreased swelling or persistent pain in bones, joints, back, or legs
L ump or mass, especially in the abdomen, neck, chest, pelvis, or armpits
D evelopment of excessive bruising, bleeding, or rash
C onstant infections
A whitish color behind the pupil
N ausea which persists or vomiting without nausea
C onstant tiredness or noticeable paleness
E ye or vision changes which occur suddenly and persist
R ecurrent or persistent fevers of unknown origin

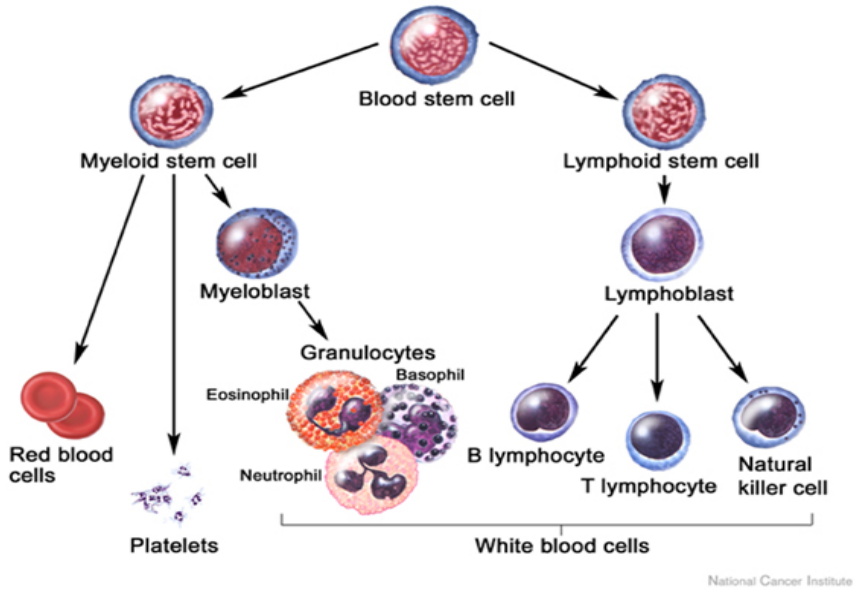
ACUTE LYMPHOBLASTIC LEUKEMIA (ALL)

ALL is the most common cancer seen in children.

WHAT IS LEUKEMIA?

Leukemia is a cancer of the blood and bone marrow. The bone marrow is the soft, spongy tissue found inside of bones where normal blood cells are made. The blood cells that are made in the bone marrow include the white blood cells (WBCs), red blood cells (RBCs), and platelets. Each type of blood cell has its own job in the body. WBCs are the infection-fighting cells. RBCs provide oxygen and energy to the body. Platelets help blood to clot; the blood's ability to clot is important during certain situations, such as when you have a cut.

Leukemia occurs as a result of abnormal growth of immature blood cells. These cells are called *blast cells*. These immature cells grow out of control, crowd-out the normal cells (WBCs, RBCs, and platelets) in the bone marrow, and eventually spill out into the bloodstream. As a result, leukemia may be found in other parts of the body such as the lymph nodes, liver, spleen, central nervous system (which is the brain and spinal cord), testicles, skin, or other organs. Leukemia occurs in cells that develop from either the lymphoid or myeloid cell lines.

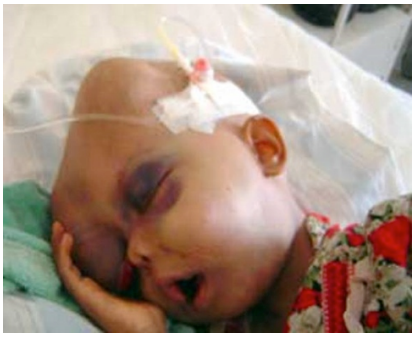


- Acute Lymphoblastic Leukemia (ALL) results from abnormalities in the lymphoid cell line
- ALL can also be called acute lymphoid, lymphoblastic, and precursor B and precursor T leukemia.
- Acute Myelogenous Leukemia (AML) results from abnormalities in the myeloid cell line – including non-lymphoid white cells, erythroid and platelets.
- Chronic Myeloid Lekemia (CML) results from an abnormality is proliferation of abnormal mature myeloid cells

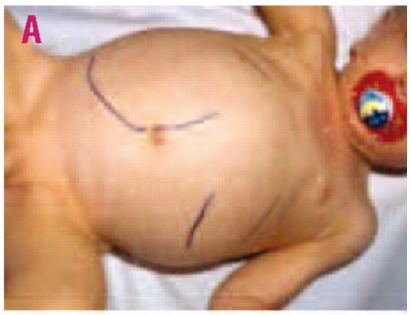
ALL PROGNOSIS FACTORS

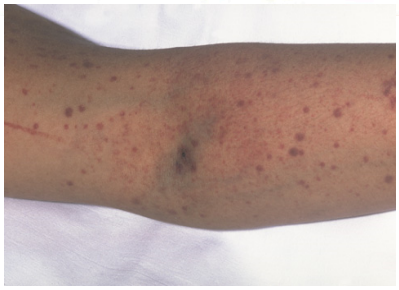
- Age at Diagnosis (children 1-10 years old are likely to have most favorable outcome)
- Initial Leukocyte Count (less than 50,000/mm is more favorable)
- Speed of response to treatment (quicker response indicates better outcome)

WHAT A PATIENT MAY LOOK LIKE AT DIAGNOSIS



- Abnormal complete blood count (CBC)
- Fever and unexplained infections
- Bruising, petechiae (small pinpoint red dots on the skin), epistaxis (nose bleeds)
- Fatigue, weakness
- Stomach pain or an enlarged stomach (due to enlarged liver and spleen)
- Pallor, anemia
- Weight loss
- Bone pain
- Lymphadenopathy (swollen/enlarged lymph nodes)





DRUGS USED TO TREAT ALL

- Induction Therapy: Usually includes weekly vincristine, a corticosteroid (such as prednisone or dexamethasone) and asparaginase; higher risk patients may also get doxorubicin or daunorubicin.
- CNS Prophylaxis (intrathecal methotrexate or methotrexate/cytarabine/hydrocortisone)
- Consolidation Therapy: aspariginase, methotrexate, vincristine, doxorubicin, corticosteroid, cytarabine, mercaptopurine
- Maintenance Therapy: Usually continues 2-3 years and includes mercaptopurine and methotrexate, usually with intermittent doses of vincristine and a corticosteroid

SPECIAL NURSING CONSIDERATIONS FOR CARING FOR A CHILD WITH ALL

- Observe for Tumor Lysis Syndrome and manage fluid balance very carefully
- Assess pain and provide pain medication and comfort as needed
- Assist with diagnostic tests (such as bone marrow aspiration and biopsy, trucut biopsies, LP and cytospin)
- Draw blood tests as needed
- Assess for signs and symptoms of infection, bleeding, and any mental status changes
- Reconstitute and administer chemotherapy as ordered
- Provide instruction on mouth care to prevent mouth sores
- Provide instruction on good nutrition
- Ensure proper hygiene and clean environment

LYMPHOMA

Hodgkin's, Non-Hodgkin's (including Burkitt's)

WHAT IS HAPPENING TO THE PATIENT

- Cancer of the lymph nodes
- Solid tumors are formed in affected lymph nodes by rapidly growing cells
- Most commonly seen in the cervical and supraclavicular (sternal) area
- Spreads to the spleen, liver and bone marrow
- Usually caused by Epstein-Barr Virus (common in children who are HIV positive)

WHAT A PATIENT MAY LOOK LIKE AT DIAGNOSIS

- Patient usually has noticeable bulge around cheek and neck, or abdomen, sometimes very large
- Chest Xray MUST be performed at diagnosis due to the increased risk of respiratory or circulatory collapse from an undiagnosed large mediastinal mass.
- Look for frequent coughing, stridor or grunting. The patient may not being able to lay flat because it is too hard to breathe.
- Early presentation: Fevers, night sweats and large amount of rapid weight loss (at least 10% of weight; for example: a child weighs 30kg prior to diagnosis and loses 3kg and now weighs 27kg)

DRUGS USED TO TREAT LYMPHOMA

- MOST COMMON: Vincristine;Cyclophosphamide; Methotrexate IV and IT (intrathecally); Doxorubicin; Cytarabine IV and IT; Bleomycin
- FOR HIGH RISK PATIENTS: Etoposide; Ifosfamide (with Mesna)

SPECIAL NURSING CONSIDERATIONS FOR CARING FOR A CHILD WITH LYMPHOMA

- Monitor for breathing problems (Trachea moved to the side) related to mediastinal mass
- Make sure the patient is urinating frequently! Get the parents to collect the urine, the child should pass at least 3mls/kg/hr. If patient passes less, it is an emergency and notify the Doctor! (See Tumor Lysis Syndrome)

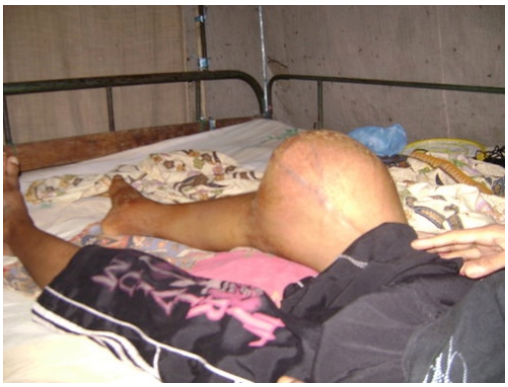
OSTEOSARCOMA

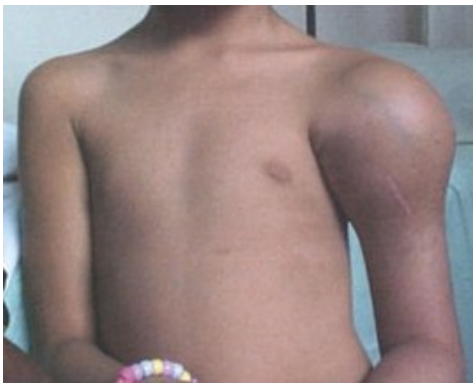
WHAT IS HAPPENING TO THE PATIENT

- Cancer of the bone. Osteosarcoma usually arises in a bone and destroys local tissue and weakens the bone.
- Because osteosarcoma usually develops from osteoblasts (the cells that make growing bone), it most commonly affects teens that are experiencing a growth spurt. It usually occurs in adolescents and young adults, but occasionally occurs in younger children.
- Osteosarcoma most often starts in the bones around the knee joint, in the upper or lower leg next to the knee, or in the thigh. The second most common place for osteosarcoma to develop is in the upper arm bone, close to the shoulder. However, osteosarcoma can develop in any bone in the body.

WHAT A PATIENT MAY LOOK LIKE AT DIAGNOSIS

- Pain in the bone or joint that gets worse over time
- A painless swelling or a noticeable mass in the arm or leg
- A broken bone that occurs without or with minimal injury or trauma
- Stiffness or swelling of joints (uncommon)
- Back pain or loss of bowel or bladder control related to a tumor in the pelvis or at the base of the spine. This is very rarely the first sign that a child has osteosarcoma.



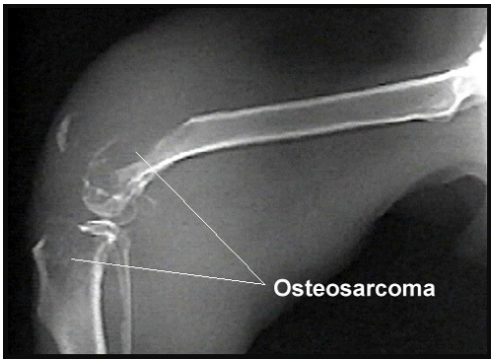


DRUGS USED TO TREAT OSTEOSARCOMA

- Cisplatin, Doxorubicin, and high dose methotrexate in some settings
- Surgery usually required to remove part of the affected limb

SPECIAL NURSING CONSIDERATIONS FOR CARING FOR A CHILD WITH OSTEOSARCOMA

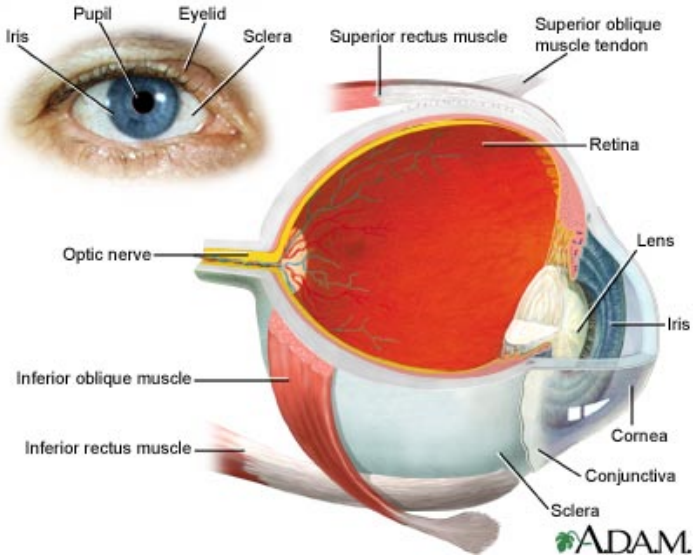
- After surgery, monitor surgical site to prevent infection
- Pain management
- Encourage rehabilitation after surgery.
- Monitor chest x-ray of lung, which is the most prevalent site for metastasis



RETINOBLASTOMA

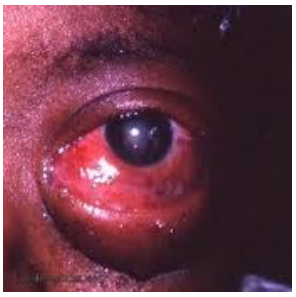
WHAT IS HAPPENING TO THE PATIENT

- Cancer of the retina of the eye, which is the thin layer of nerve tissue that coats the back of the eye
- Occurs most often in children under the age of 4
- Can occur in one or both eyes



WHAT A PATIENT MAY LOOK LIKE AT DIAGNOSIS

- A pupil that looks white or red instead of the usual black- on careful questioning a white pupil is almost always noted at some point in the child's history
- A crossed eye (looking toward the nose or the ear)
- Poor vision
- A red, painful eye
- An enlarged pupil
- Differently colored irises
- Children with metastatic disease may be inconsolably crying, unable to walk, or have swelling distant to the affected eye.

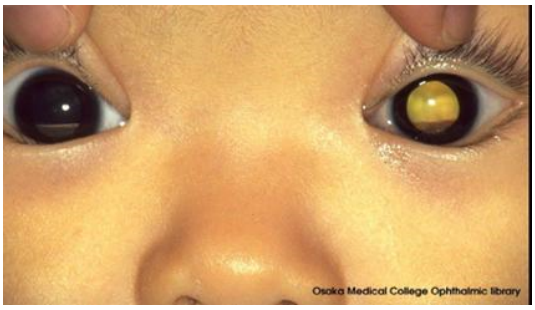


DRUGS USED TO TREAT RETINOBLASTOMA

- Vincristine, Carboplatin, Etoposide, cyclophosphamide, doxorubicin, Cytarabine IT
- Surgery almost always required in resource poor settings to remove the affected eye

SPECIAL NURSING CONSIDERATIONS FOR CARING FOR A CHILD WITH RETINOBLASTOMA

- Monitor for anaphylaxis during Carboplatin/etoposide administration, can happen within minutes of infusion (see anaphylaxis information sheet)
- Monitor for vision changes and signs of infection in eye

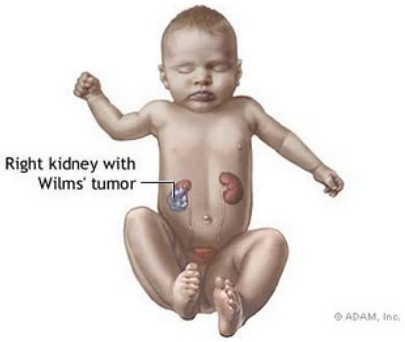


WILM'S TUMOR

Tumor of the Kidneys

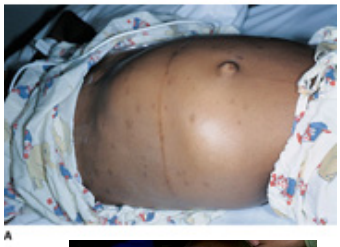
WHAT IS HAPPENING TO THE PATIENT

- Tumor of one kidney, or both kidneys
- Has four stages of severity
  1. Stage I: Tumor confined to one kidney – able to treat only with surgery
  2. Stage II: Tumor extends outside of the kidney but can be completely removed surgically
  3. Stage III: Small or large amounts of tumor remain after surgery – including around the abdominal cavity or into lymph nodes
  4. Stage IV: Tumor has metastasized outside of the abdominal cavity into the surrounding lymph nodes, or into the lung, liver, or rarely bones or brain
  5. Stage V: Tumors in both kidneys at diagnosis
- Treated with a combination of chemotherapy, surgery, and radiation.



WHAT THE PATIENT MAY LOOK LIKE AT DIAGNOSIS

- Patient will have small or large abdominal mass. Could be one-sided or two-sided depending on stage of disease
- Possible renal complications (for example: decreased urine output or NO output) depending on size of tumor

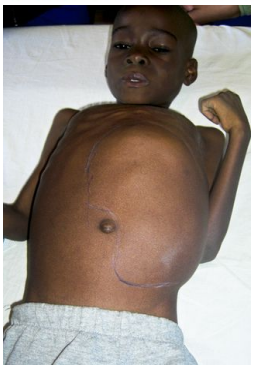


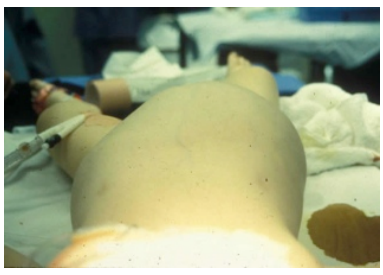
DRUGS USED TO TREAT WILM'S TUMOR

- MOST COMMON: Actinomycin, Vincristine
- FOR HIGH-RISK (HIGHER STAGED) PATIENTS: Doxorubicin, Cyclophosphamide, Etoposide

SPECIAL NURSING CONSIDERATIONS FOR CARING FOR A CHILD WITH WILM'S TUMOR

- !!!!!!!!!DO NOT PALPATE THE ABDOMEN!!!!!!! The tumor breaks apart VERY easily!! If it is palpated – it can break apart and spread to other parts of the body very quickly!
- Biopsies should be done at the time of surgery once the tumor is removed, not with needle biopsy
- After surgery, the patient will only have one kidney because that is the only way to remove the tumor. Monitor intake and output carefully to make sure the patient is not being fluid overloaded.
- Be very careful when using nephrotoxic antibiotics, for example gentamicin or vancomycin. Check blood renal function prior to starting these.





Nursing Implications for Patients with Neuroblastoma
Oncopedia #438

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Added to Oncopedia: 01/19/2011

Introduction

Neuroblastoma arises in the sympathetic nervous system, and is the most common extracranial solid tumor of childhood, with between 600 and 700 new cases diagnosed each year in the United States.[1-3] Neuroblastoma is one of several solid tumors that occurs only in children, and very rarely develops in children over 10 years of age or in adults.[1,4] It is estimated that 2-5% of patients have a familial predisposition to developing neuroblastoma.[3] The prognosis for neuroblastoma patients is dependent on age, stage, site of primary tumor, pattern of metastases, and genetic markers; survival rates range from 40% to 90%. [2] Proper nursing management of these patients is imperative and consists of thorough patient assessments, knowledge of administration guidelines for treatment, and patient and family education.

Management Concerns for the Nurse

1. Patient Assessment

Clinical manifestations of neuroblastoma are dependent upon the location of the primary tumor and/or metastatic disease.[1-4] Primary tumors manifest as abdominal tumors, pelvic tumors, thoracic tumors, or cervical tumors. Seventy percent of all neuroblastoma tumors arise in the abdomen.[4] Therefore, a thorough physical assessment of the abdomen as well as a detailed history of any abdominal pain, fullness, or discomfort is imperative for proper nursing management of patients with neuroblastioma. Primary pelvic tumors may cause bowel and/or bladder dysfunction; again, a detailed history is imperative in order to determine the severity of the disease.

Patients with thoracic tumors may present with dyspnea and increased difficulty breathing. Any difficult breathing or changes in respiratory status should immediately be reported to the healthcare team.[4] Neuroblastoma may also present as a cervical tumor and can manifest as a palpable mass, Horner's syndrome, heterochromia (a range of colors) in the iris, and rarely, superior vena cava syndrome.[4] The Oncopedia document *Head and neck neuroblastoma. Clinical presentation simulating cervical adenopathy*,[7] gives an example of neuroblastoma presenting as a cervical mass.

Symptoms of metastatic disease depend on where the metastases occur. Sites of metastasis are bone, bone marrow, liver, lungs, brain and soft tissue.[1] It is crucial to remember that two-thirds of neuroblastoma patients present with metastases to the bones, bone marrow, lymph nodes, liver, or subcutaneous tissue.[3] Metastasis in the brain and lungs are rare and are only seen in association with end-stage disease or a relapse. [4] Some other characteristic clinical findings in neuroblastoma patients are periorbital ecchymosis, Pepper syndrome, and blueberry muffin syndrome. The Oncopedia chapter on *Neuroblastic Tumors* provides more details on common characteristics of patients with neuroblastic tumors. The Oncopedia videos *Head and Neck Neuroblastoma: Clinical Presentation as an Upper Respiratory Obstruction*[5] and *Neuroblastoma 4S*[6] show some presenting features of neuroblastoma

Like other childhood cancers, a detailed patient history is essential in the nursing care of children with neuroblastoma. This history should include (a) if, when, and how a palpable mass was discovered; (b) any other clinical manifestations that have presented; (c) any family history of neuroblastoma.[3]

2. Treatment Administration

Treatments for neuroblastoma differ based on the staging of the tumor. Within the International Neuroblastoma Staging System (INSS), tumor classifications range from stage 1 (localized tumor confined to the area of origin) to stage 4 (dissemination of tumor to distant lymph nodes, bone, bone marrow, liver, and/or other organs – except those organs defined in stage 4S). Notably, patients with the MYCN amplification are always considered to have a worse prognosis than patients without this genetic abnormality.[3]

Surgery alone is used to treat patients with stage 1 or stage 2 disease. Surgery with chemotherapy and radiotherapy is used it to treat patients with stage 3 and stage 4S disease. Patients with stage 4 disease are treated with much more aggressive chemotherapy, radiation, and surgery. Autologous stem cell rescue is also used to treat these patients.[3]

Table 1*

Chemotherapeutic agents commonly used in the treatment of neuroblastoma.

Chemotherapy	Common Side Effects	Other Administration Notes
Carboplatin	Very intense nausea and vomiting, electrolyte imbalance	**Risk of anaphylaxis even after first few doses are given**
Cyclophosphamide	Nausea, vomiting, bone marrow suppression	May cause **hemorrhagic cystitis**
Doxorubicin	Cardiac dysfunction, bone marrow suppression, nausea, vomiting	**Vesicant;** Will cause red-colored urine; May need cardiac testing prior to administration
Etoposide	Hypotension Weakness	**Risk of anaphylaxis** **Vigilant blood pressure monitoring**
Irinotecan	Diarrhea; Abdominal cramping Sweating Fever	**Diarrhea/abdominal cramping usually treated with Atropine**

*Includes information from the *Lippincott Manual of Nursing Practice*. [2]

As always, the comfort of the patient with neuroblastoma is a high priority for nursing staff. Analgesics should be given for pain control. Antiemetics may be given for nausea and vomiting and antipyretics may be used for the treatment of fever. Intravenous fluids may be administered to ensure an acceptable fluid balance in these patients, and antibiotics may be given to prevent or control any postoperative infections.

3. Staging Neuroblastoma

Recently, in an effort to allow for comparison of risk-based clinical trials, the International Neuroblastoma Risk Group (INRG) classification system was developed to establish a consensus approach for pretreatment risk stratification. For this initiative, the statistical and clinical significance of 13 potential prognostic factors were analysed in a cohort of 8,800 children who were diagnosed with neuroblastoma between 1990 and 2002. The cohort was composed of children from North America and Australia (Children's Oncology Group), Europe (International Society of Pediatric Oncology Europe Neuroblastoma Group and German Pediatric and Hematology Group), and Japan. A complete description of this staging system can be found in the Oncopedia chapter titled *Neuroblastic Tumors.* [3]

4. Patient and Family Education

Patient and family education for neuroblastoma patients and their families consists primarily of diagnostic and treatment information. Due to the vast array of treatments that may be used based upon the disease stage, many tests are performed at the time of diagnosis to ensure the disease is staged accurately. Education on the different tests that will be performed, including laboratory tests, diagnostic imaging, and potential treatments, is absolutely essential in order to decrease anxiety in these patients and their families. Of special note, a 24-hour urine is usually collected on these patients (as 95% of all neuroblastoma tumors secrete catecholamines in the urine); therefore, proper education on collection of the 24-hour urine is vital.[1] Infection control precautions and immediate treatment of fever are also key elements of patient and family education.

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Nursing Implications for Patients with Rhabdomyosarcoma
Oncopedia #598

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Added to Oncopedia: 12/23/2013

Introduction

Rhabdomyosarcoma (RMS) in children is a soft tissue tumor that is thought to arise from mesenchymal cells.[i] However, cells that give rise to RMS have not been clearly identified, as tumor cells can appear both in or around muscle beds and in areas of the body that do not have skeletal muscle, such as the genitourinary tract.[1] The most common areas where RMS occurs are in the head, neck, and genitourinary tract, with a small percentage (20%) in the extremities.[2] RMS is categorized as either embryonal (the most common and generally found in young children) or alveolar and anaplastic RMS; it may also be categorized as an undifferentiated sarcoma is more common in the extremities and trunk in adolescents.[3,4] Ultimately, RMS can develop anywhere in a child's body except the bone[2] and generally presents with pain and symptoms related to the specific area of the body where the tumor is located. Most children (~80%) are diagnosed by 14 years of age; however, there are peaks in incidence at the ages of 2-4 years and 12-16 years.[5] An Iranian study showed that children with localized tumors had an ~80% survival rate for stages I and II,[6] while another recent study in Morocco found an overall survival rate of 70% (event free survival, 39%) for children with RMS, although 37% of the children abandoned treatment.[7] Overall, in high-income countries, more than 70% of children with non-metastatic RMS survive their disease for at least 5 years.[5] Histologically, the 2 distinct subtypes of RMS, embryonal and alveolar, have specific genetic alterations. Alveolar RMS can be confirmed by polymerase chain reaction (PCR) showing fusion genes, and the tumors are normally located in extremities, while head and neck tumors are generally embryonal (**Table 1**).

Table 1
Features of RMS by subtype and location[2]

Rhabdomyosarcomas		
Subtype	***Location***	***Symptoms***
Alveolar • Confirmed by PCR • Fusion genes • Small, round, densely packed cells lined up along spaces reminiscent of pulmonary alveoli (6)	Extremities with high rate of regional lymph node metastases	• Relatively fixed to the underlying musculature and occasionally involve the skin[8]
Embryonal • Approximately 35% of RMS • Rarely spread to lymph nodes • Spindle-shaped cells, stromal-rich appearance	**Orbital** tumors	• Proptosis and sometimes ophthalmoplegia
	Parameningeal tumors	• Nasal, aural, or sinus obstructions can have mucopurulent or sanguineous discharge • If it extends into the cranium, it can cause cranial nerve palsy or meningeal symptoms
	Other head and neck tumors	• Generally painless, growing mass that is localized
Genitourinary		
	Bladder	• Bladder tumors cause hematuria and obstruction of the urinary tract
	Prostate	• Large pelvic mass causing urinary problems (compression of the bladder) or constipation (compression of the intestinal tract)
	Vaginal	• Vaginal tumors (generally in very young children) have a grape-like appearance—*botryoides* • Mass or vaginal bleeding or vaginal discharge, but rare for regional lymph node involvement
	Uterine	• Usually older female patients and quite extensive by diagnosis
	Paratesticular	• High rate of lymph node metastases to the retroperitoneum (particularly in male patients >10 years of age)
Alveolar • Confirmed by PCR • Fusion genes • Small, round, densely packed cells lined up along spaces reminiscent of pulmonary alveoli [6]	Extremities with high rate of regional lymph node metastases	• Relatively fixed to the underlying musculature and occasionally involve the skin (8)

Histologic subtypes of RMS are divided according to the modified International Classification of Rhabdomyosarcoma. The categories and expected survival, in parentheses, are as follows: spindle cell (very good), botryoid (95%), embryonal (60%), and alveolar (54%).[8]

Metastasis can include bone involvement with symptoms that resemble bone tumors or leukemia, including pain, limping, or swelling. If the metastasis is in the bone marrow, symptoms can include pancytopenia (bleeding, anemia, and/or infection).[8]

Management Concerns for the Nurse

1. Patient Assessment

Pre-operative workup of RMS includes laboratory work and imaging studies, including computer tomography or magnetic resonance imaging. For metastatic disease, a bone marrow aspirate, bone scan, CT of the brain, lungs, and liver, as well as lumbar puncture for the cerebrospinal fluid collection are performed.[9] The imaging is important to determine if surgical resection is possible, and if neo-adjuvant therapy will be necessary to diminish the size of the tumor. Regional and distant lymph node evaluation is also required for pre-treatment staging,[9] and the tumor's histological subtype and size as well as the child's age will significantly determine the outcome of therapy.[10] **Table 2** describes the TNM staging criteria; in the US, the former grouping system of staging is used for radiation therapy (see **Table 4** under *Surgery*).

Table 2*
Intergroup Rhabdomyosarcoma Study Group Staging System based on a modified TNM (tumor, node, metastasis)[8]

			Description		
Stage	**Site**	**T**	**Size**	**N**	**M**
1	Orbit Head and Neck Genitourinary	T_1 or T_2	a or b	N_0, N_1, or N_x	M_0
2	Bladder/Prostate Extremity Cranial parameningeal Other	T_1 or T_2	a	N_0, or N_x	M_0
3	Bladder/Prostate Extremity Cranial parameningeal Other	T_1 or T_2	a b	N_1 N_0, N_1, or N_x	M_0 M_0
4	All	T_1 or T_2	a or b	N_0, or N_1	M_1

Abbreviations: a, less than or equal to 5 cm; b, greater than 5 cm in diameter; M, metastasis; M_0, no distant metastasis; M_1, metastasis present; N, regional node; N_0, regional nodes not clinically involved; N_1, regional nodes clinically involved; N_x, status of regional nodes unknown; T, tumor invasiveness; T_1, tumor confined to site of origin; T_2, extension and/or fixation of tumor to the surrounding tissue.

* Adapted from Kotsubo CZ. Rhadomyosarcoma. In: Baggott CR, Fochtman D, Foley GV, Kelly KP, editors. Nursing Care of Children and Adolescents with Cancer. 4th ed. Glenview, IL: Association of Pediatric Hematology/Oncology Nurses; 2011. p. 1060.

2. Treatment Administration

2.1 *Chemotherapy*

Chemotherapeutic agents commonly used to treat RMS are shown in Table 3. Since the majority of children with RMS are assumed to have micrometastasis, all children receive chemotherapy. A 3-drug combination of vincristine, dactinomycin, and cyclophosphamide (VAC) has been used in North America, and recent trials have shown that the addition of ifosfamide, etoposide, or topotecan did not result in significant improvement in the clinical outcome.[11] However, in Europe, clinical trials including anthracyclines and alkylating drugs (e.g., ifosfamide) have been conducted to attempt to avoid radiation therapy in some cases.[11]

Table 3
Chemotherapeutic agents commonly used in the treatment of RMS

Chemotherapy	Common Side Effects	Other Administration Notes
Vincristine (12)	• nausea and vomiting • stomach pain and cramps • constipation • diarrhea • jaw pain, headache, or other aches • thinned or brittle hair	Vincristine is a vesicant; thus, leakage into surrounding tissue can cause significant damage. Determining adequate blood return before, during and following administration of vincristine is essential. If extravasation occurs, follow extravasation protocols immediately.[14]
Dactinomycin [13]	• vomiting • stomach pain • diarrhea • hair loss	Gastrointestinal toxicity and marrow suppression occurs following therapy with both dactinomycin and radiation. The buccal and pharyngeal mucosa can have early erythema. Erythema caused by earlier radiation may reactivate when the patient receives dactinomycin alone.[15]
Cyclosphosphamide[16]	• nausea • vomiting • loss of appetite or weight • abdominal pain • diarrhea • hair loss • sores on the mouth or tongue • changes in skin color • changes in color or growth of finger or toe nails	This medication is best taken early in the morning, so significant hydration can occur during therapy and for two days after therapy. Irritation of the bladder can cause hemorrhagic cystitis so the patient may require MESNA and IV fluids to prevent this irritation. The drug can also cause changes in taste. If high doses of cyclophosphamide are to be given, child may receive an echocardiogram before and during treatment to monitor heart function. The drug can cause fertility problems in the future. Taking this medication simultaneously with itraconazole, fluconazole, or ketoconazole can enhance the side effects of the medication.[17,18]

Ifosfamide[19]	• Bladder irritation • Potential liver damage • Can cause rash or dry skin and sensitivity to sun	Must drink at least two liters a day and may receive Mesna to protect bladder Sunscreen recommended

There is considerable philosophical disparity in RMS treatment approaches (e.g., use of radiation versus surgery for local therapy) for RMS between European and US cooperative groups.[10,20] Since RMS is a rare disease, children who are eligible for clinical trials are limited, and only long-term follow-up will truly reveal the late-effects of treatment; thus, the outcome of treatment philosophies will take years to determine.[10] Research on the tumor biology of RMS will be essential for future improvements in therapy as well as a decrease in the substantial compromising health issues that many survivors endure.

RMS "tumor-specific fusion proteins" could be ideal therapeutic targets although current evidence suggests that for most targeted therapies, combinations of drugs might be needed to achieve improvements in survival rates and to prevent the rapid appearance of resistant clones".[10] RMS includes a diversity of tumors, each of which responds distinctively to treatment; therefore, improved diagnosis, classification and more tumor-specific treatment that reduces late effects will be the challenge for the future.

2.2 *Radiation*

Radiation therapy can be divided into two categories: brachytherapy—"delivered from within or by contact with the use of an implant or mold with radioactive sources" or teletherapy—"techniques using an external radiation source delivered at a distance from the patient".[21] In a study by the Children's Oncology Group (D9602), in the US, it was found that lowered radiation doses provided local control in patients with low-risk embryonal RMS if cyclophosphamide was included in their systemic therapy.[21] In the US, children with very low-risk embryonal tumors that are totally resected do not receive radiation therapy. Those with intermediate-risk tumors receive a lower than standard dose (36 Gy) and those with high-risk (and unresectable) tumors receive 50.4 Gy.[8] In Europe, local control is restricted to surgery and does not include radiation therapy.[20]

Important considerations for children and adolescents receiving radiation therapy include their continuing physical growth and development, compliance for young children,

and late toxicities for patients who have received radiotherapy.[21] Two-thirds of children with RMS are less than six years old.[9] Toxicities can include damage to lung or liver, salivary glands, thyroid glands, decreased fertility, heart problems, impaired neurocognition, and ototoxicity. Ultimately, a second neoplasm is most worrisome. In one study among survivors of soft tissue sarcomas, 8.8% had a second neoplasm and in girls, 23% of these cancers were breast cancers.[21]

2.3 *Surgery*

An open biopsy of a child's mass is used to confirm RMS. Core needle biopsies are less invasive, but result in reduced tissue samples, increase errors in sampling and inconclusive findings, and preclude the option of molecular studies.[9] Pretreatment re-excision is a wider surgery with adequate margins of normal tissue performed prior to adjuvant therapy; the importance of adequate margins has been shown to result in better outcomes than either gross or even microscopic residual tumor.[9] Second-look surgeries (most effective for children with trunk or extremity tumors) have been used to verify the tumor's response to therapy. "The goal of second-look surgery is complete resection of partial responders or non-responders." [8] There is no consensus at the moment about surgery for children with a residual mass after treatment; thus, in the US, current practice is to follow the tumor with serial imaging.[8] For a table of the clinical grouping system used to guide surgery planning, see **Table 4**.

Table 4*
Clinical Grouping System

Group I	Localized disease, completely resected A. Confirmed to organ or muscle of origin B. Infiltration outside organ or muscle of origin; regional nodes not involved
Group II	Compromised or regional resection including A. Grossly resected tumors with microscopic residual tumor B. Regional disease, completely resected, with nodes involved and/or tumor extension into an adjacent organ C. Regional disease, with involved nodes, grossly resected, but with evidence of microscopic residual tumor
Group III	Incomplete resection or biopsy with gross residual disease remaining
Group IV	Distant metastases present at onset

**Adapted from Leaphart C, Rodeberg D. Pediatric surgical oncology: management of rhabdomyosarcoma. Surg Oncol 2007 Nov;16(3):177.*

2.4 *Poor Tumor Response*

Patients with metastatic RMS generally have a poor prognosis. Unfavorable factors include "time to relapse, metastatic recurrence, prior radiation therapy, and the size of the initial tumor (>5 cm)".[11] In general, if possible, surgical resection and radiation therapy for sites that have not been irradiated previously are recommended for local control. However, salvage chemotherapy treatment is not universally defined and previous treatment guides chemotherapy after relapse.[11] In a study in Iran, the five-year survival rate for children whose disease had recurred was 31.91%; [6] however, in general only approximately 25% of children who have relapsed will survive after five years.[8] Since bone marrow transplant has not been shown to be effective for these children, new therapeutic strategies are being studied that include "rebeccamycin, termozolomide, vinorelbine and docetaxel".[8] Whether a child's relapse is local or systemic does not seem to affect ultimate survival.[8]

3. Patient and Family Education

Understanding the complexity of RMS diagnosis, staging, treatment plans and lifetime follow-up is not simple. The timing of laboratory testing, imaging, surgery, radiation therapy, and possible second surgeries during treatment can challenge even the most sophisticated families. Therefore, nurses are uniquely positioned to support patients and families with consistent and regular review and explanations of the treatment plan. Patients and families who do not speak the local language of the hospital or treatment facility and those with low levels of education or income are particularly vulnerable for misunderstanding or not fully understanding a complex treatment plan. Nurses must make an extra effort to be sure these patients and families are clear about the entire treatment process and any locally available resources (both economic and psychosocial).

Nurses can also educate families about expected side effects from all three forms of treatment (surgery, chemotherapy, and radiation) to raise the patient and family's awareness of what to anticipate and the options in place to manage these symptoms. For example, in one study in the US, almost 1/5 patients with intermediate risk RMS had a >10% weight loss while receiving treatment.[22] Although this weight loss did not result in a lower survival rate, it did increase toxicity; thus, vigilant efforts to maintain nutrition in children

and adolescents with RMS is important and a vital nursing focus in concert with registered dieticians, if available.

In a study of children with various cancers, including RMS, in Jordan, parents noted that the burden of caring for their child with cancer was more than they had expected. Two groups of parents were interviewed: newly diagnosed children's parents and those whose child had been diagnosed 6-12 months earlier. Less than half the parents described themselves as "at peace with themselves and their situation in life".[23] Parents whose children were newly diagnosed had higher levels of anticipatory grieving, as did parents who were from low-income families.[23] Thus, nurses must be prepared to support the parents of children with RMS (as well as other cancers), particularly in the early period following diagnosis.

The intensity of chemotherapy and the expected side effects of all treatment for RMS imply that the nursing role for symptom management will be challenging. Consistent communication with the patient and family about a child or adolescent's individual response to treatment will allow for accommodations or interventions that may decrease suffering and discomfort.

Finally, the significant late effects of treatment for RMS demand that children and adolescents who survive their disease must be followed by an adult physician who is familiar with pediatric cancer (and in this case, RMS) treatment. Ongoing surveillance of the former patient's physical, mental, spiritual and emotional health is paramount to a successful adulthood. Nurses are uniquely qualified to ensure that this transition takes place since they often see former patients over time, either in the community or for initial follow-up visits in pediatric oncology units.

Notes

i. "An embryonic connective tissue cell with an outstanding capacity for proliferation and capable of further differentiation into reticular cells or osteoblasts". (http://medical-dictionary.thefreedictionary.com/mesenchymal+cell accessed May 5, 2013)

Resources

Sarcoma Alliance in California, US
http://sarcomaalliance.org/what-you-need-to-know/sarcoma-specific-information/

Sarcoma Foundation of America in Maryland, US
http://www.curesarcoma.org/index.php/patient_resources/

The Liddy Shriver Sarcoma Initiative in New York, US
http://sarcomahelp.org/index.html

Sarcoma Patients EuroNet in Riemerling, Germany
http://www.sarcoma-patients.eu/

Asociación Española de Afectados por Sarcomas (AEAS) in Madrid, Spain
http://www.aeasarcomas.org/

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ACTINOMYCIN-D

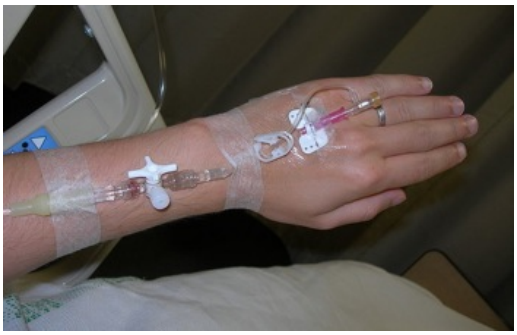
Dactinomycin, Cosmegan

USE

Chemotherapy medication used for the treatment of Wilms Tumor, Ewings Sarcoma, Rhabdomyosarcoma

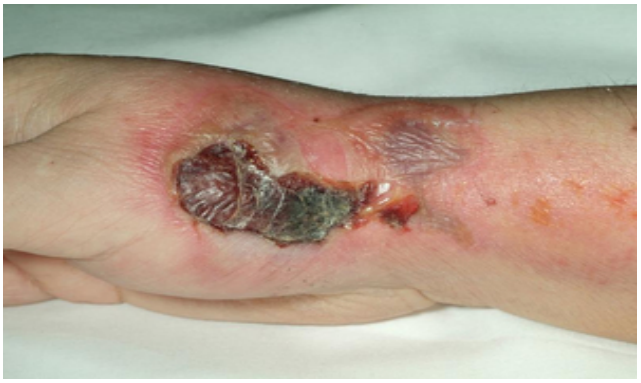
ADMINISTRATION (IV)

- Clear yellow liquid administered intravenously
- Dilute with NS and administer over 10-15 minutes



POSSIBLE SIDE EFFECTS

- Nausea and vomiting
- Loss of appetite
- Increased risk of infection, anemia, bleeding (due to decreased in white blood cells, red blood cells, and platelets)
- Hair loss
- Mouth sores
- Acne
- Skin Rash or redness
- Skin sensitivity to sunlight
- Veno-occlusive disease-manifests as abdominal pain and hepatomegaly



NURSING IMPLICATIONS

- Premedicate with antiemetics such as ondansetron, granisetron or diphenhydramine
- Actinomycin is a VESICANT; SEVERE tissue damage can occur with extravasation.
  - Cold Compresses should be applied if extravasation occurs
- Protect medication from light
- Inhalation of dust or vapors and contact with skin or mucous membranes, especially those of the eyes, must be avoided. Always wear protective equipment when handling.
- Do not use a filter on the IV tubing as this may remove some of the medication.
- Instruct patient to protect skin from sunlight when outdoors (wear long clothing)

ASPARAGINASE (Elspar, L-asparaginase)
PEG-ASPARAGINASE (Pegaspargase, Oncaspar)

USE

- Chemotherapy medication used for the treatment of Leukemia
- Asparaginase is an enzyme made from the bacteria *escherichia coli* (E. coli).
- PEG-linked asparaginase (or PEG-Asparaginase) is a slightly changed version of the E. coli form linked to polyethylene glycol (PEG) molecule which makes the drug stay in the body longer.
- PEG-Asparaginase may be given if patient has shown hypersensitivity reaction to another form of Asparaginase, however the risk of reaction would be higher (per the insert).

ADMINISTRATION (IM/Intramuscular, IV)

- PEG-Asparaginase is given as an infusion into a vein or an injection into a large muscle
- For IM injection: each injection should be <2ml, so sometimes multiple injections are needed and should use 2 different sites per injection
- For IV infusion: give over 1-2 hours and monitor for signs of allergic reaction

POSSIBLE SIDE EFFECTS

- Local allergic reaction is common
- Systemic allergic reaction characterized by wheezing, difficulty breathing, swelling, hives, itching, rash, irritation
- Anaphylaxis is possible; THIS IS A MEDICAL EMERGENCY; see Anaphylaxis information sheet; Anaphylaxis can be days after administration as drug is slowly released into body. Incidence of anaphylaxis may increase with subsequent doses.
- More immediate reaction would be expected with the IV dose
- Fever, aches, chills, and elevated liver function labs
- Nausea, Vomiting
- Rash
- Poor appetite; stomach pain
- Has been associated with pancreatitis
- Lethargy and somnolence are the most common symptoms of CNS toxicity.
- Hyperglycemia or an increase in blood sugar
- Has been associated with coagulopathy or a defect in body's ability to form blood clots correctly; this may result in excess bleeding or clotting

NURSING IMPLICATIONS

- Have emergency medications (epinephrine, diphenhydramine, steroids), oxygen, and resuscitative equipment available
- Observe patient closely for at least 60 minutes after dose
- Alert family members that anaphylaxis can occur days later and is a MEDICAL EMERGENCY
- Asparaginase is contraindicated for patients with a history of pancreatitis and who have had significant hemorrhagic events with past asparaginase therapy.

BLEOMYCIN

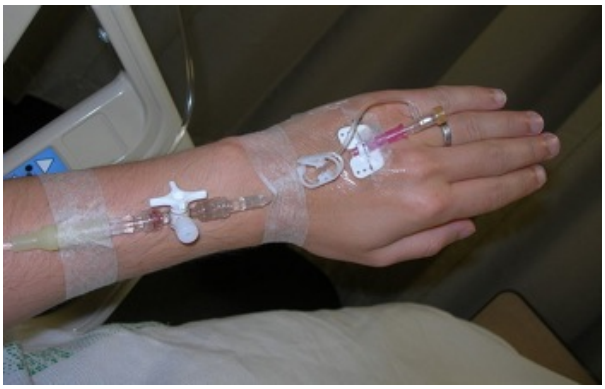
USE

Chemotherapy medication used to treat Hodgkin's Lymphoma

ADMINISTRATION

- Comes as a powder in the vial
- Add 5ml of NS to vial, the concentration would then be 3 units/ml.
- IV infusion over 10 minutes

POSSIBLE SIDE EFFECTS



- Hair loss
- Hyperpigmentation of fingernails
- Mouth sores
- Anaphylaxis (fever, low blood pressure, difficulty breathing)
- Pneumonitis, pulmonary fibrosis (long term side effect)
- Nausea, vomiting
- Rare: High fevers 2-6 hours after administration, believed to be from the release of pyrogenic cytokines in sensitive individuals with lymphoma

NURSING IMPLICATIONS

- Monitor patient during test dose for an allergic reaction. Monitor blood pressure, temperature and for difficulty breathing (if this occurs, refer to handout on anaphylaxis)
- Anti-nausea medications as available such as ondansetron, granisetron, or diphenhydramine to prevent vomiting
- Inspect for mouth sores and encourage good oral hygiene

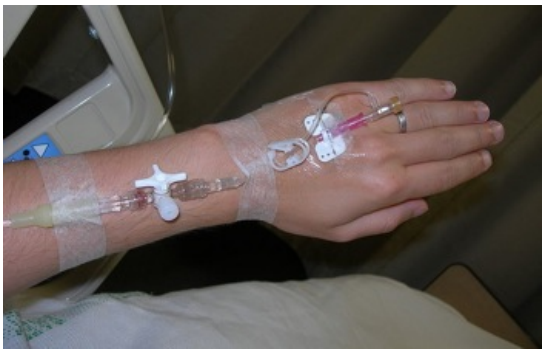
CARBOPLATIN

USE

Chemotherapy medication used for the treatment of Retinoblastoma and Wilm's Tumor

ADMINISTRATION

- Vial comes as a powder, do not refrigerate vials
- Must inject D5W, 0.9% NaCl, or sterile water to the vial to make concentration 10mg/ml (So if it is a 50mg vial, inject 5ml and the concentration would be 10mg/ml)
- Then you would draw up the dose you need
- After you draw up the correct dose, further dilute with 250ml of D5W or 0.9% NaCl
- Infuse over 30min through peripheral IV
- Do not use any aluminum needles or IV tubing, if medication comes in contact with aluminum it will cause precipitation and the medication will not be effective



POSSIBLE SIDE EFFECTS

- Nausea, vomiting usually beginning 6 hours after and could last up to 24 hours
- Increased risk of infection, anemia, bleeding (due to decreased in white blood cells, red blood cells, and platelets)
- ANAPHYLAXIS, risk of allergic reaction increases with each dose given
- Electrolyte imbalance
- Metallic taste

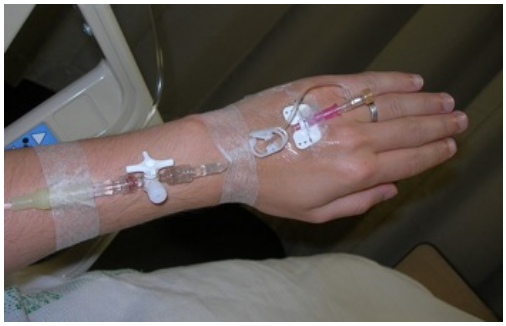
NURSING IMPLICATIONS

- Premedicate with antiemetics such as ondansetron, granisetron, or diphenhydramine.
- Continue to medicate for nausea as needed
- Monitor for symptoms of anaphylaxis with each dose, symptoms such as rash, hives, itching, difficulty breathing, fever

CISPLATIN

USE
Chemotherapy medication used for the treatment of Osteosarcoma

ADMINISTRATION (1mg/ml concentration)



- After you draw up ordered dose, dilute with 250-500ml of 0.9% NaCl
- Administer through a peripheral IV over 60 minutes
- Protect the medication from light
- Do not refrigerate vials
- Do not use any aluminum needles or IV tubing, if medication comes in contact with aluminum it will cause precipitation and the medication will not be effective
- Depending on dosage, the rate can be anywhere from 60 minutes up to 8 hours

POSSIBLE SIDE EFFECTS

- SEVERE nausea and vomiting
- Increased risk of infection, anemia, bleeding (due to decreased in white blood cells, red blood cells, and platelets)
- Low levels of magnesium
- Hearing loss
- Metallic taste
- Electrolyte imbalance
- Anaphylaxis (see anaphylaxis sheet)
- Nephrotoxicity
- Neurotoxicity

NURSING IMPLICATIONS

- Premedicate with antiemetics such as ondansetron, granisetron, or diphenhydramine
- Continue to medicate for nausea at least 24hours after therapy
- HYDRATION is very important, give IV fluids per protocol and monitor intake and output, encourage oral hydration
- Monitor hydration status such as skin turgor, mucous membranes, tears, and impact of nausea and vomiting on oral intake
- Maintain urinary output of at least 2ml/kg/hr, if they aren't maintaining good urine output, may need to increase IVF or give a 0.9% Sodium Chloride bolus
- Supplement with magnesium if necessary

CYCLOPHOSPHAMIDE

Cytoxan

USE

Chemotherapy medication used for the treatment of Osteosarcoma, High-risk Wilm's tumor, Leukemia, and Lymphoma

ADMINISTRATION

- 500mg, 200mg vials, 1 gram vials, 2 gram vials
- Draw up ordered dose and dilute it to 20mg/ml with 0.9% Sodium Chloride or D5W (usually around 50-250ml)
- Administer through a peripheral IV over 30-60 minutes
- Give dose after a 0.9% Sodium Chloride bolus (2 times maintenance IV rate) and follow medication with 2-4 hours of 0.9% Sodium Chloride fluid (1-1.5 times maintenance IV rate)

POSSIBLE SIDE EFFECTS

- Nausea and vomiting
- Myelosuppression (decrease in blood counts, such as white blood cells, red blood cells, and platelets
- SIADH (syndrome of inappropriate antidiuretic hormone hypersecretion) meaning that the patient can have low sodium levels and become fluid overloaded
- Aim for a specific gravity of 1.010 or less
- Decreased appetite (anorexia)
- Loss of hair
- Reproductive sterility
- Bleeding of the bladder

NURSING IMPLICATIONS

- Premedicate with nausea medications such as diphenhydramine, granisetron or ondansetron
- !!!HYDRATION both before and after the medication is very important to prevent the bladder from bleeding!!!
- Maintain adequate hydration and urinary output
- Check urine frequently for blood and specific gravity. Try to get a specific gravity of urine that is 1.010 or lower
- Give the medication early in the day so the toxic side effects to the bladder do not accumulate overnight
- Encourage patient to void before going to bed at night
- Administer slowly (30-60 minutes) to avoid an itchy nose, numbness around the mouth and lightheadedness

CYTARABINE

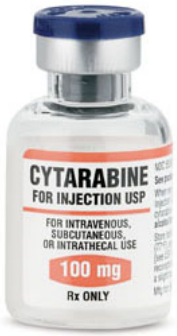
Ara-C, Cytosar, Cytosine Arabinoside

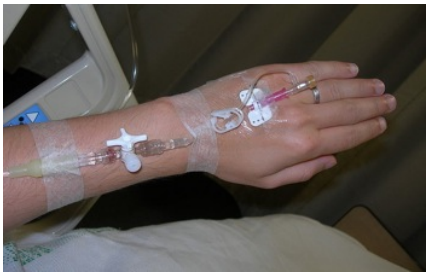
USE

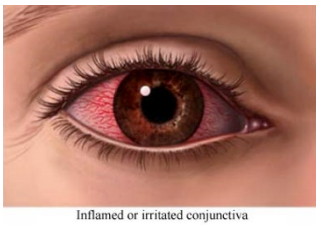
Chemotherapy medication used for the treatment of Leukemia, Lymphoma

ADMINISTRATION (IV, IM, SQ, IT)

- Dilute with 0.9% NaCl or D5W
- Administer through a peripheral IV over ordered time (may be 1-2 hours, or continuous infusion over 24 hours)



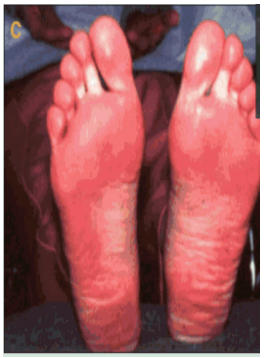




Inflamed or irritated conjunctiva

POSSIBLE SIDE EFFECTS

- Nausea and vomiting
- Loss of appetite
- Conjunctivitis (“Pink Eye”) or inflammation of the outermost layer of the eye and the inner surface of the eyelids may occur with high dose (greater than 2 grams) Cytarabine
    - If this occurs, patient may be particularly sensitive to light and may wish to cover their eyes with a washcloth or sunglasses
- Increased risk of infection, anemia, bleeding (due to decreased in white blood cells, red blood cells, and platelets)
- Hair loss
- Flu-like symptoms: fever, aches, fatigue
- Cerebellar toxicity
- Rash
- Peeling of skin on hands and feet may also occur with high dose therapy



NURSING IMPLICATIONS

- Premedicate with antiemetics such as ondansetron, granisetron, or diphenhydramine
- Steroid Eye Drops or artificial tear drops may be needed with high doses of Cytarabine to prevent or treat conjunctivitis
- Continue to medicate for nausea at least 24 hours after therapy

DACARBAZINE

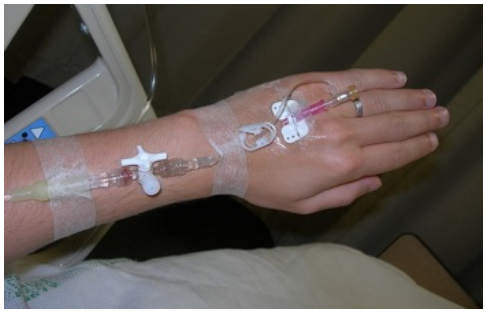
DTIC

USE

Chemotherapy medication used for the treatment of Hodgkin lymphoma and sarcoma

ADMINISTRATION (IV)

- Clear liquid given into a vein
- Store the vials in the refrigerator, away from light
- Reconstitute with sterile water for injection and then dilute with either D5W or NS for infusion



POSSIBLE SIDE EFFECTS

- Moderate to severe nausea and vomiting
- Loss of appetite
- Increased risk of infection, anemia, bleeding (due to decreased in white blood cells, red blood cells, and platelets) 2 to 4 weeks after treatment
- Flu-like symptoms: fever, aches, fatigue
- Metal taste in mouth during infusion

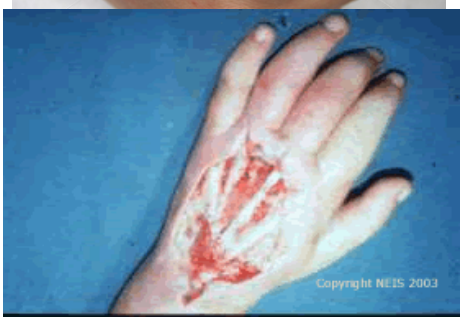
NURSING IMPLICATIONS

- Premedicate with antiemetics such as ondansetron, granisetron, or diphenhydramine
- Continue to medicate for nausea at least 24 hours after therapy
- Protect from light
- Dacarbazine is an irritant and can harm tissues if it leaks out of the vein
    - If this occurs, apply warm compresses and protect the tissue from light for 48 hours

DOXORUBICIN (ADRIAMYCIN)
DAUNORUBICIN

USE
Chemotherapy medication used to treat Leukemia, Lymphoma, and Wilms tumor

ADMINISTRATION (2mg/ml) concentration



- Vials of Doxorubicin
- Infusion through IV
- Extravasation burn from Doxorubicin
- Red in color
- Dilute with 50ml of 0.9% Sodium Chloride
- Give medication through IV infusion over 15minutes, or 30 minutes or 24 hours depending on dose and protocol (protect the medication from light)
- Vesicant, meaning if the medicine leaks outside the vein, it can cause painful tissue damage and blistering. Only give through properly placed IV. Must be able to draw back blood from the IV to make sure the IV is in the vein properly. Monitor for redness, swelling, or pain at IV site to prevent extravasations.

POSSIBLE SIDE EFFECTS

- Cardiac arrhythmias and toxicities can be seen during the doxo/dauno infusion especially if the patient is on a cardiac monitor. The long-term cardiac toxicities manifest more like congestive heart failure.
- Nausea and vomiting
- Pink or red color to the urine
- Increased risk of infection, anemia, bleeding (due to decreased in white blood cells, red blood cells, and platelets)
- Ulcers in mouth
- Hair loss
- Rare: anaphylaxis, rash (see Anaphylaxis sheet for instructions)

NURSING IMPLICATIONS

- Anti-nausea medications as available such as ondansetron, granisetron, or diphenhydramine to prevent vomiting
- Explain to parents that urine may be red in color for 1-2 days
- Inspect for mouth sores and encourage good oral hygiene
- Warn the parents that some or all of the child's hair may fall out slowly, but it is temporary

ETOPOSIDE

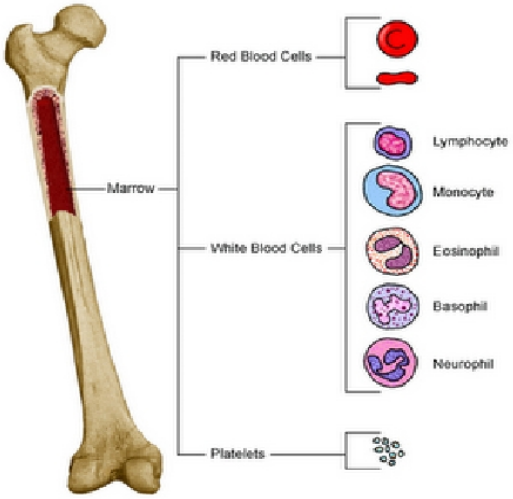
VP-16

USE

Chemotherapy medication used for the treatment of Retinoblastoma, Lymphoma and high-risk Wilm's Tumor

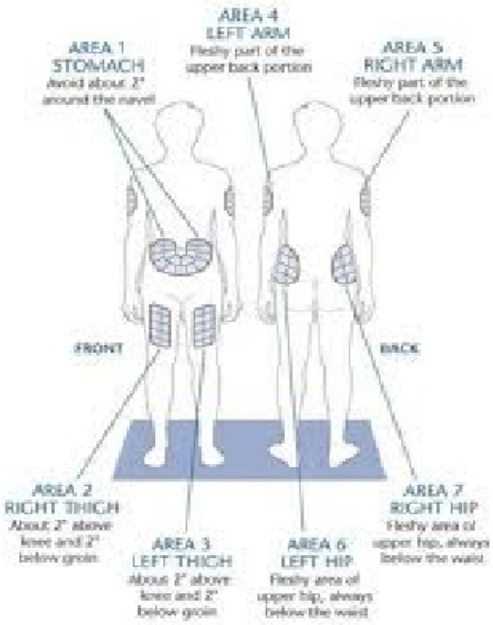
ADMINISTRATION (Available in 100mg vials)

- Administer drug SLOWLY!!! Can drop the patient's blood pressure very low if given too quickly
- Mixed with 0.9% Sodium Chloride and given at LEAST over 1 hour through a peripheral IV
  - 1mg-100mg in 250ml 0.9% NaCl
  - 101mg-200mg in 500ml 0.9% NaCl
- Avoid a concentration above 0.4mg/ml
- Can administer over 2 hours if the patient is smaller or younger to prevent low blood pressure
- Check for precipitates in the fluid (white crystals)
- DO NOT REFRIGERATE!!



POSSIBLE SIDE EFFECTS

- Hypotension (low blood pressure) not so common when a 100mg/m2 doses are given over one hour
- Nausea and vomiting
- Increased risk of infection due to lower white blood cell count
- Numbness and tingling of fingers or toes
- Mouth sores
- Secondary cancers



NURSING IMPLICATIONS

- Avoid rapid infusion to prevent low blood pressure
- Monitor blood pressure frequently during infusion if possible (every 15 minutes)
- Instruct patient to change positions slowly (Blood pressure drops faster when patient moves from laying down to sitting too quickly)
- Premedicate with antiemetics such as ondansetron, granisetron, or diphenhydramine to prevent nausea

FILGRASTIM

GCSF

USE

Medication used to boost the immune system after chemotherapy administration in solid tumor patients only - NOT chemotherapy

ADMINISTRATION

- Subcutaneous injection is the preferred route (although some give it IV. The drug is more rapidly eliminated with IV doses so the response to the same dose could be less).
- Do not shake vials
- Any vial or syringe in room temperature for more than 24 hours should be thrown away - make sure drug is refrigerated!!!
- Subcutaneous Injection Sites
- Filgrastim stimulates bone marrow to produce White Blood Cells

POSSIBLE SIDE EFFECTS

- Bone pain
- Joint pain
- Pain at injection site
- Rare: anaphylaxis

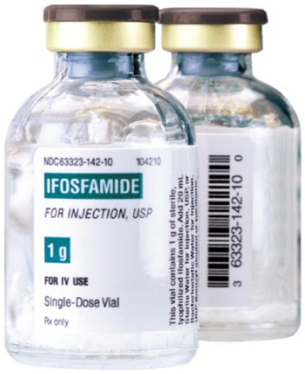
NURSING IMPLICATIONS

- Instruct patient that lower back and legs might ache for several days after drug is given
- Use hot pack on sore areas
- Monitor patient for one hour after FIRST dose to assess for anaphylactic reaction

IFOSFAMIDE (Ifex)
MESNA (Mesnex)

USE

Ifosfamide is a chemotherapy medication used for the treatment of various cancers including lymphomas, sarcomas, and bone tumors. It is similar to cyclophosphamide. Mesna is a “chemoprotectant” or “antidote” and is used to prevent the harmful side effect of Ifosfamide known as hemorrhagic cystitis or bleeding and irritation of the bladder.

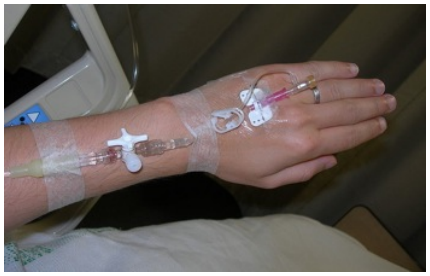


ADMINISTRATION (Ifosfamide = IV) (Mesna = IV or PO)

- Ifosfamide is a white powder that gets mixed with sterile water to form a clear liquid for IV infusion
- Administer through a peripheral IV over ordered time

POSSIBLE SIDE EFFECTS

- Nausea and vomiting
- Loss of appetite
- Increased risk of infection, anemia, bleeding (due to decreased in white blood cells, red blood cells, and platelets)
- Hair loss
- Neurotoxicity: sleepiness, confusion, hallucinations
- Risk of severe hemorrhagic cystitis if given without mesna
    - Mesna may be mixed with Ifosfamide or may be given separately.
    - IV dose may be given orally at a higher dose but has a bad taste and children may not like it.
    - Mesna works by binding with the main metabolite or byproduct of Ifosfamide (acrolein) so it cannot harm or irritate the bladder lining.



NURSING IMPLICATIONS

- Premedicate with antiemetics such as ondansetron, granisetron, or diphenhydramine
- Monitor fluid intake and output
- Hydration must begin 3-6 hours before Ifosfamide dose and continue 24 hours after dose
- MESNA MUST BE GIVEN EXACTLY AS ORDERED AND ON TIME
- Frequently check urine for blood. Blood may be present but not easily seen or may be clearly seen (pink or red urine)!
- Monitor concentration of urine (known as urine specific gravity); goal is specific gravity (s.g.) less than 1.010

MERCAPTOPURINE

6-MP, Purinethol, 6-Mercaptopurine

Please note: The name of the drug is 6-MP. It does NOT mean to give 6 of the pills.

USE

Oral chemotherapy medication used for the treatment of Leukemia, Lymphoma

ADMINISTRATION (PO)

- Usually given during maintenance phase
- Store in a dry place at room temperature
- Dose should be taken daily at one time, preferably at bedtime on an empty stomach (2 hours after eating)
- Milk and grapefruit juice decrease absorption so do NOT give with these drinks.
- If patient is also taking allopurinol (a medication for tumor lysis syndrome), the dose of mercaptopurine must be reduced

POSSIBLE SIDE EFFECTS

- Myelosupression or decreased bone marrow activity results in increased risk of infection, anemia, bleeding (due to decreased in white blood cells, red blood cells, and platelets)
- Mercaptopurine dose may need to be held/reduced if myelosupression is extreme
- Decreased appetite
- Nausea, vomiting
- Diarrhea
- Mucositis
- Darkening of skin
- Liver toxicity including hyperbilirubinemia

NURSING IMPLICATIONS

- Make sure dose is given at same time each day, 2 hours before/after eating
- Pills may need to be cut depending on ordered dosage
- Encourage patient to drink plenty of fluids while on treatment

METHOTREXATE

USE

Chemotherapy medication used for the treatment of Leukemia and Lymphoma

ADMINISTRATION

- Is administered orally, by IV or into the spinal fluid
- Draw up ordered IV dose and dilute with 0.9% Sodium Chloride (see chart below)
    - 150mg-499mg = 50ml 0.9% NaCl over 30 minutes
    - 500mg-1500mg = 250ml 0.9% NaCl over 60 minutes
    - Greater than 1500mg = 1000ml 0.9% NaCl over 1-6 hours
- Protect drug from light
- Use preservative free saline to mix with the methotrexate when giving it into the spinal fluid

POSSIBLE SIDE EFFECTS

- Nausea and vomiting (can be severe if administered too quickly)
- Decreased appetite (anorexia)
- Mouth sores
- Increased sensitivity to the sun
- Increased bilirubin (jaundiced - yellow eyes/skin) - damage to the liver
- Peeling/redness to palms of hands and soles of feet
- Mental status changes (toxicity)
- Spinal fluid administration side effects: nausea, headache, dizziness, learning disabilities

NURSING IMPLICATIONS

- Premedicate with antiemetics such as diphenhydramine, ondansetron or granisetron
- Do not give the patient their sulfamethoxazole and trimethoprim (Bactrim/Septra) while receiving IV Methotrexate (sulfamethoxazole and trimethoprim compete with Methotrexate for secretion in the renal tubules and can increase Methotrexate toxicity)
- Patients should not receive non-steroidal anti-inflammatory agents just before or after Methotrexate due to increased toxicity.
- Monitor liver enzymes if possible - check for increased jaundice both to skin and eyes both during and after drug is administered
- Only doses lower than 100mg can be given IV push - others need to be given more slowly to avoid severe nausea and vomiting
- Hydration and/or hydration with sodium bicarbonate (to increase urine alkalinization) are often used with higher doses
- Patients will need prophylactic Leucovorin rescue for higher doses of IV Methotrexate. Leucovorin is a medication given either IV or PO to decrease the harmful effects of Methotrexate.

PREDNISONE
DEXAMETHASONE

USE

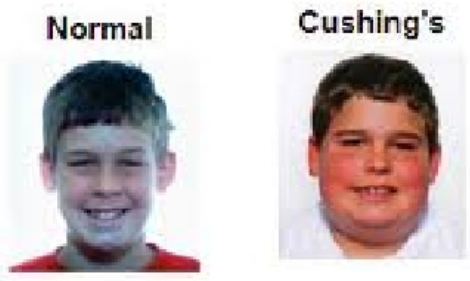
Steroids used for the treatment of Leukemia. Can also be used for nausea management and to achieve an anti-inflammatory response

ADMINISTRATION (Available in varying mg tabs)

- To be taken with food
- If possible, do not crush as medication tastes very bitter
- Can mix with a small amount of food to hide the taste
- Co-administer with ranitidine (Zantac) if available to avoid abdominal upset

POSSIBLE SIDE EFFECTS

- High blood glucose (sugar) level
- Personality changes (patients become very angry and irritable)
- Weight gain (cheeks appear larger= Cushing's Syndrome)
- Increased appetite
- Acne (Pimples)
- Abdominal Upset (heartburn)
- Increased risk of infection (low white blood cell counts)

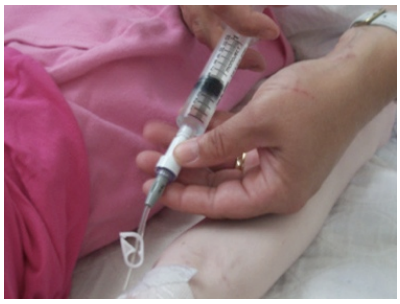


NURSING IMPLICATIONS

- May prevent a fever in the patient even if the patient has an infection (see Fever and Neutropenia sheet).
- Monitor the patient for other signs of illness: irritability, unable to wake patient up, cough/cold symptoms
- Monitor for high blood sugar levels: frequent urination, increased thirst, possible muscle cramping
- Provide patient with frequent, small meals - reducing salt intake if possible
- Always give medication on a full stomach

VINCRISTINE
VINBLASTINE

DO NOT GIVE VINCRISTINE THROUGH THE SPINE



Picture of giving Vincristine through IV

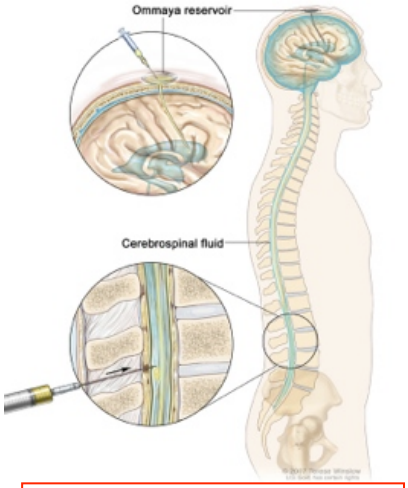
Picture of a burn from Vincristine if it leaks outside the IV

USE

Chemotherapy medication used for treatment of leukemia, lymphoma, and Wilm's tumor.

ADMINISTRATION (1mg/ml concentration)

- Max dose of Vincristine is 2mg (no matter what the weight of the child is)
- Push medication through the peripheral IV over 2-5 minutes
- Vesicant, meaning if the medicine leaks outside the vein, it can cause painful tissue damage and blistering. Only give through properly placed IV. Must be able to draw back blood from the IV to make sure the IV is in the vein properly. Monitor for redness, swelling, or pain at IV site to prevent extravasations.
- FATAL if given intrathecally (into the spine), only give through the IV. Do not even bring vincristine into the room of a patient getting intrathecal medication.



Picture of intrathecal injection into the spine

POSSIBLE SIDE EFFECTS

- Jaw pain (infants may have difficulty sucking, monitor for adequate feeding)
- Constipation (may need increased fiber in diet or stool softener)
- Numbness and tingling of hands/feet
- Hair loss (warn the parents that some or all of the child's hair may fall out slowly, but it is temporary)
- Increased risk of infection, anemia, bleeding (due to decreased in white blood cells, red blood cells, and platelets)

NURSING IMPLICATIONS

- Notify doctor if patient has numbness and tingling of hands/feet, difficulty walking, drooping of eyelids
- Explain the importance of good hand washing and keep child away from other sick children
- Vinblastine may cause a decrease in hemoglobin and platelet levels, Vincristine does not!
- Vincristine doesn't affect the bone marrow, but Vinblastin does!
- Vincristine can cause neurotoxicity, but Vinblastin does not.
- Beware of the drug interaction with "azole" antifungals which will greatly increase the toxicity of Vincristine and Vinblastine.
    - Pay particular attention to prophylactic antifungals (such as itraconazole) that many countries give during induction therapy.

DRUG EXTRAVASATIONS

Definition

Escape of intravenously infused chemotherapeutic agents outside the vein into the surrounding tissues either due to leakage from the vein. Chemotherapy drugs can be categorized as vesicants or irritants based on their potential to cause tissue damage.

- A vesicant drug is one that causes blister formation with tissue necrosis and subsequent sloughing of dead tissues
- Irritants are drugs capable of causing inflammation and irritation but they rarely produce any tissue breakdown

Risk factors for extravasation

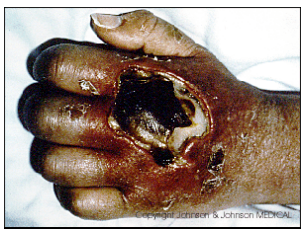
- Poor technique and inexperience of person placing the IV
- Poorly secured needle with dislodgement of the needle tip into surrounding tissues
- Inappropriate selection of the position and size of IV and the length of time for which the IV is left in place

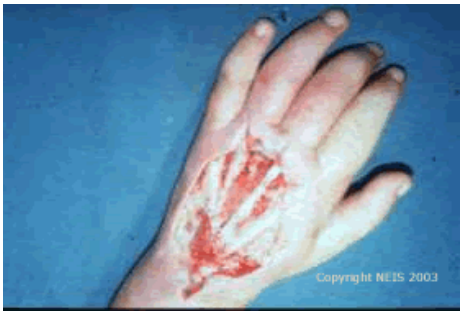
Preventive measures

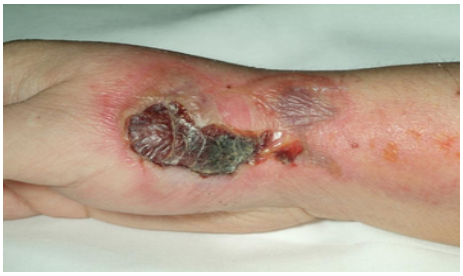
- Allow only authorized practitioners who have been adequately trained in chemotherapy administration to give chemotherapy
- Administer cytotoxics through a recently placed IV. Tape the IV so it cannot become dislodged. Many institutions require that the IV site be “fresh”, meaning less than 24hours old
- Take particular care in the selection and positioning of the IV. Use the forearm and avoid, if possible, IV sites near joints
- IV should be securely taped to prevent “in and out” motions which can lead to the IV coming out completely and/or to an enlargement of the hole in the vein leading to extravasations of medication(s) despite the fact that the IV is still in place and blood can be aspirated

Signs and symptoms

- Pain, stinging, burning or any other acute change at the injection site.
- Redness, swelling, or leakage at the injection site
- Increased resistance to administration
- No blood return is obtained. If this is found, other signs should be verified, as this can be very misleading because there can still be extravasations with blood return.







DRUG EXTRAVASATIONS (continued)

Monitoring

The most important step in dealing with extravasations is prevention. Ensure that all drugs are administered by appropriately trained personnel who are familiar intravenous access devices, the drugs being administered and common irritants and vesicants. They should also be capable of identifying and acting upon the first signs of extravasation.

Drug Extravasations Management

1. **Stop the infusion, leave the needle in place and attempt to draw back any remaining drug from the IV.**

2. **Apply warm or cold pack. See chart to determine which one to use. Avoid applying pressure. Elevate the extremity with the affected area.**
   - COLD compresses help stop spread of the drug and prevent further injury. Vasoconstriction caused by cold helps localize the extravasation.
   - WARM compresses causes vasodilation and increases systemic absorption and distribution of the chemotherapy. Warm compresses reduce potential for tissue damage. HOWEVER, this is true ONLY for vinca alkaloids. Where indicated, apply firmly but without undue pressure a heat source (hot water bottle or small electrically heated blanket) to the area continuously for 24 hours. The heat source should not be in direct contact with the skin and a piece of dry gauze should be laid in between. This assists the natural dispersal of the drug.

3. **Notify the Doctor and monitor for increased pain, swelling, and change in color.**

4. **Drugs**
   - Hyaluronidase
     This is an effective antidote for vinca alkaloids. 150 units of hyaluronidase is diluted in 2 ml of water for injection, or 0.9% Sodium Chloride. Multiple subcutaneous injections and then gentle massage of the affected area causes dilution of the chemotherapy and reduces tissue injury.
   - Topical Dimethyl Sulfoxide
     Recommended application for topical solution is 3 to 4 times a day. . Contact with good skin should be avoided. If blistering occurs discontinue use and seek further advice.
   - Steroids
     Topical hydrocortisone 1% can potentially reduce non-specific inflammation. So it is still recommended for use in non-vinca alkaloid injuries.

5. **Surgical intervention**
   - Excision Wide excision with use of grafts may be indicated if persistent pain 1-2 weeks after injury. Inadequate excision is associated with continuing necrosis at the margins, poor granulation and failure of engraftment.

Drug Extravasations

Table 1. Specific antidotes in the management of peripheral extravasation

Drug/class of drug	Warm/Cold compression	Specific antidote
Vinca Alkaloids Vincristine Vinblastine Vinorelbine	**WARM** compression – apply for 24 hours	**Hyaluronidase 150 IU** Draw up 150 IU hyaluronidase in 1 to 2ml water for injection or 0.9% Sodium Chloride. Inject 0.1 to 0.2ml subcutaneously at points of the compass around the circumference of the area of extravasation. Gently massage area to facilitate dispersal.
Daunorubicin Doxorubicin Actinomycin-D (Dactinomycin) Dacarbazine	Apply **COLD** pack intermittently for 30 minutes in every 2 hours for 24 hours. Place a piece of dry gauze between skin and cold pack	**Topical dimethyl sulfoxide (DMSO) 50%** Apply Topical DMSO 50% using a cotton bud or with your gloved hand. Avoid contact with good skin. For the next 7 days apply DMSO50% every 6 hours alternating with topical hydrocortisone 1% cream every 3 hours. Do not use an occlusive cover. If blistering occurs, stop DMSO and seek further advice.
Cisplatin Carboplatin Etoposide All other chemo	Automatic cold or warm compression is **not** required. However if symptoms warrant then use intermittent cold compression except in the case of cisplatin, or carboplatin when warm or cold compression may be used.	**No specific antidote needed** If signs of erythema persist then **topical 1% hydrocortisone cream** may be used. Apply sparingly to the affected area 4 times a day

NAUSEA / VOMITING

Nausea and vomiting are common side effects of chemotherapy treatment for cancer. But in most cases, these side effects can be controlled with preventive medications.

Acute	Delayed	Anticipatory
▪ Begins within several hours of chemotherapy and produces the most severe symptoms during the first 12-24 hours ▪ Usually self-limited	▪ Begins 24 hours after chemotherapy has been administered ▪ Can last for up to 2 weeks	▪ Begins before chemotherapy has been given or during first hours ▪ Risk factors include inadequate control of previous nausea and vomiting, age, pre-existing anxiety or depression

MEDICATION MANAGEMENT

Symptom management: Prevention is important. Give medications prior to chemotherapy to help prevent nausea. Then continue to give as needed after the chemotherapy. You can give more than 1 anti-nausea medication if it is needed.

- Ondansetron (Zofran) can be given every 4-6 hours as needed in a dose of 0.15 mg/kg q4h x 3 or 0.45 mg/kg once daily…used best before giving chemotherapy to prevent nausea instead of once nausea has started. Parenteral doses should be given as a short infusion rather than IV push due to known EKG changes.
- Granisetron (Kytril) can be given twice a day, usually every 12 hours as needed or cab be given once daily
- Diphenhydramine (Benadryl) can be given every 6 hours as needed
- Lorazapam (Ativan) can be given every 4-6 hours as needed
- Promethazine (Phenergan) can be given every 6 hours as needed

NAUSEA / VOMITING (continued)

COMMON CAUSATIVE AGENTS

High-risk (>90% without anti-nausea)	Moderate (30-90%)	Low/minimal (<30%)
▪ Cisplatin ▪ Cyclophosphamide (>1.5 g/m^2) ▪ Dactinomycin ▪ Dacarbazine	▪ Cytarabine (>1 g/m2) ▪ Cyclophosphamide (<1.5 g/m2) ▪ Doxorubicin ▪ Daunomycin ▪ Carboplatin ▪ Ifosfamide ▪ Irinotecan	▪ L-Asparaginase ▪ Cytarabine (<1 g/m2) ▪ Bleomycin ▪ Vincristine ▪ Vinblastine ▪ Methotrexate (<100 mg/m^2) ▪ Etoposide ▪ Methotrexate (> 100 mg/m^2) ▪ 5-FU ▪ Cytarabine (<100 mg/m^2)

OTHER POSSIBLE, NON-CHEMO RELATED CAUSES TO CONSIDER

- Acute gastroenteritis
- Anticipatory/psychogenic – adolescents, females, previous experience
- Gastric ulcer disease
- Hepatitis
- Increased intracranial pressure
- Intestinal obstruction, which may be related to narcotic use for pain control
- Urinary tract infection

GENERAL CONSIDERATIONS

Counseling, nutritional & hydration status, patient/family education regarding nausea/vomiting and preventive measures that are used

ASSESSMENT

Potential adverse outcomes from nausea & vomiting include dehydration, electrolyte imbalance and malnutrition

- Weight, nutritional status including appetite, frequency, hydration status
- Medication/disease history

MONITORING

Weight, nutritional status, frequency, hydration status (vitals, physical exam), Intake and Output balance, electrolytes (Na(sodium), K(potassium), Glucose)

FEVER & NEUTROPENIA

Information taken from **INCTR Supportive Care Handbook** at http://inctr.wikidot.com, which was produced by the **INCTR Pediatric Oncology Strategy Group**.

Infections represent one of the most important causes of morbidity and mortality in patients treated with chemotherapy.

DEFINITIONS

Fever

- Single oral temperature of > 38.3°C (101°F), or
- A temperature of 38.0°C (100.4°F) for > 1 hour

Neutropenia

Neutropenia is an abnormally low count of neutrophils, white blood cells that help your immune system fight off infections, particularly of bacteria and fungi. The lower your neutrophil count, the more vulnerable you are to infectious diseases. The **BEST** way to prevent infection in children that are neutropenic is **GOOD HANDWASHING** for **EVERYONE** involved in taking care of the child, including the parent and child themselves.

- A neutrophil count of < 500 cells/mm^3, or
- A count of < 1000 cells/mm^3 with a predicted decrease to <500 cells/mm^3

Who qualifies for initial antibiotic therapy?

- All neutropenic patients at the onset of fever
- Afebrile (without fever), neutropenic patients with signs/symptoms compatible with infection

EVALUATION OF THE NEUTROPENIC PATIENT

Clinical examination

- Signs and symptoms of inflammation (pain, redness, warmth, pus) may be minimal or absent due to neutropenia
- Sites that deserve attention during the physical exam
    - Oral cavity and pharynx
    - Lungs
    - Perineum, including the anus
    - Skin, including the bone marrow aspiration sites, intravenous catheter exit site and tissue around the nails
- Poor nutrition can add additional risk for infection

Laboratory investigations

- CBC= Complete Blood Count
- Liver function and renal function tests
- At least one set of blood cultures from peripheral vein
- Thick smear for malarial parasites (if indicated)

In settings where blood culture is not drawn administer antibiotics to complete 7 days without fever

- Chest x-ray if patient has signs/symptoms of respiratory infection

FEVER & NEUTROPENIA (continued)

MANAGEMENT

Give broad spectrum antibiotics as soon as possible after fever occurs. **Ceftriaxone** is the first drug of choice. Intravenous antibiotic choices include the following:

- **Cephalosporins**: cefuroxime, cefotaxime, ceftriaxone, ceftazidime
- **Penicillins**: ampicillin, piperacillin/tazobactam, flucloxacillin, cloxacillin
- **Aminoglycosides**: gentamicin, amikacin
- **Antifungals**: fluconazole, ketoconazole, amphotericin B

First line regimens should include a cephalosporin +/- aminoglycoside or a penicillin + aminoglycoside. For example:

- Ceftriaxone + gentamicin
- Ampicillin + gentamicin
- Ceftazidime or cefotaxime monotherapy
- Piperacillin/Tazobactam
- Where clinically indicated and available, metronidazole can be added to these regimens.
- Antifungal therapy ought to be initiated if blood cultures are negative and fever persists for five days.
- Where clinically indicated, addition of anti-viral therapy should be considered (e.g. herpes or varicella infection) using acyclovir IV or PO as available.

USE OF COLONY STIMULATING FACTORS

The administration of Filgrastim may decrease the incidence and duration of febrile neutropenic episodes. If used, a colony stimulating factor may be withdrawn once the neutrophil count is stabilized at > 500-1000/mm^3.

ANTIBIOTIC PROPHYLAXIS

Trimethoprim and sulfamethoxazole (Bactrim) prophylaxis has been proven to be effective in Pnemocystis carinii pneumonia in both neutropenic and non neutropenic patients on chemotherapy. Fungal infections are often difficult to diagnose and treat, so giving trimethoprim and sulfamethoxazole (Bactrim) to prevent fungal infections can be very beneficial.

NURSING ASSESSMENT AND INTERVENTIONS

- Obtain baseline vital signs (temperature, blood pressure, heart rate, respiratory rate, and capillary refill) and monitor frequently if an infection is suspected in a neutropenic patient
- Assess for any pain and record location and intensity
- Obtain a detailed history including:
    - Onset and duration of fever
    - Assess if any medications or treatments have been given for fever
    - Obtain list of medications patient is currently receiving or has recently received, including date of last chemotherapy treatment
    - Assess if nausea, vomiting, diarrhea are present (Patient may be dehydrated)
    - Assess for cough, shortness of breath, runny nose, difficulty breathing

FEVER & NEUTROPENIA (continued)

NURSING ASSESSMENT AND INTERVENTIONS (continued)

- Perform head to toe assessment as appropriate with attention to any areas of complaint or areas of possible entryway for bacteria
- Obtain labs and cultures as ordered

NURSING PRINCIPLES OF ANTIBIOTIC THERAPY

- Know if patient has any baseline allergies
- Double check ordered dose with formulary
- Inform and educate patient and family regarding plan of care
- Administer medication promptly as ordered
- Monitor for allergic reactions to antimicrobial treatment
- Monitor for drug interactions (especially with Vincristine and azole antifungals)
- Monitor vital signs and appearance of child closely; alert doctor to any changes
    - Septic shock(see reference sheet for Septic Shock) may be indicated by a drop in blood pressure, increase in heart rate, increase in respiratory rate, prolonged capillary refill (>3 seconds), decreased pulse ox level, or confusion/change in level of consciousness
    - Parents and caregivers often know their child best and may recognize subtle changes before the health care team does; request that they report any abnormalities as soon as possible

SUMMARY

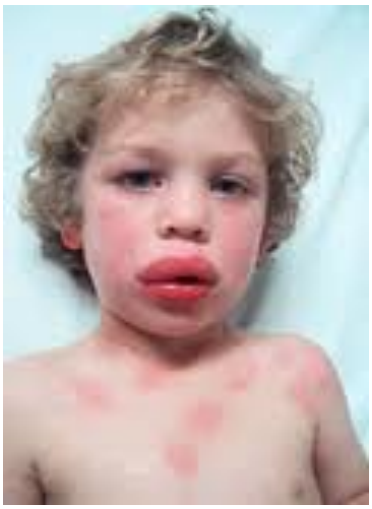
Febrile neutropenia is a common complication of cancer therapy

- Early identification
- Prompt intervention with appropriate antibiotic therapy
- Improved survival

PREVENTION IS THE KEY
KEEP AWAY FROM SICK PEOPLE AND GOOD HANDWASHING

ANAPHYLAXIS

Anaphylaxis is a rapidly progressing, life-threatening systemic (whole body) allergic reaction. Unlike other allergic reactions, however, anaphylaxis can be deadly. Reaction may begin within minutes or even seconds of exposure, and rapidly progress to cause airway constriction, skin and intestinal irritation, and altered heart rhythms. In severe cases, it can result in complete airway obstruction, shock, and death.

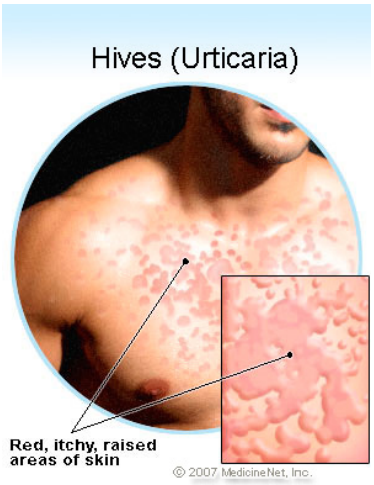


SIGNS AND SYMPTOMS

- Rash/Hives
- Difficulty breathing/Shortness of breath/Stridor/Wheezing
- Swelling in the lips, neck throat, around the eyes
- Itching
- Agitation/anxiety
- Abdominal pain
- Low blood pressure, dizziness

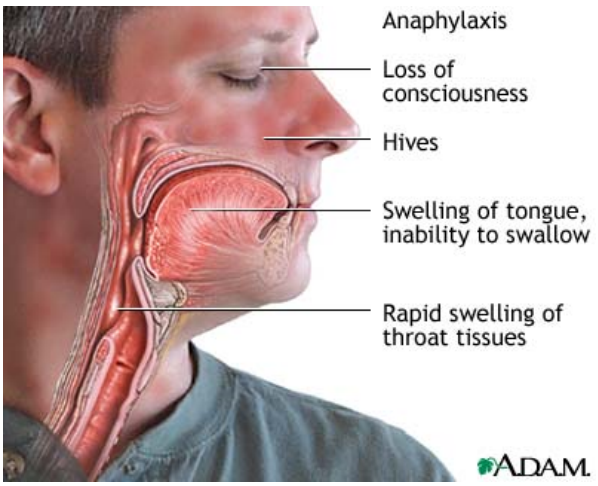
BEFORE ADMINISTERING CHEMOTHERAPY

- Get baseline vital signs prior to giving medication
- Know your patients weight (so that in an emergency you can give the right amount of the emergency medications)
- Have emergency equipment available, such as oxygen, suction, emergency medications



DURING ADMINISTRATION

- If signs and symptoms of anaphylaxis appear, STOP the infusion
- Unhook the medication from the IV and remove any medication that might still be in the IV
- Notify the doctor or other nurses to get more help
- Maintain airway, give oxygen
- Monitor Vital signs for fever, low blood pressure, tachycardia
- Medications that may need to be given:
    - Diphenhydramine
    - Epinephrine
    - Methylprednisolone
    - Ranitidine IV
    - Albuterol nebulizer
    - 0.9% NaCl bolus



SEPTIC SHOCK

MOST COMMON CAUSE OF DEATH RELATED TO TREATMENT

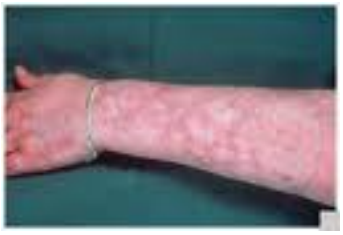
WHAT IS HAPPENING TO THE PATIENT

- Patient has a very severe infection and sometimes shows no symptoms
  - Infection from stomach virus, respiratory virus, open wounds/sores (for example, in the mouth)
- Condition can worsen very quickly (sometimes within minutes) from no symptoms to death
- USUALLY 7-10 DAYS AFTER CHEMO COMPLETE. At this time, the patient's ability to fight any infection is very low. All the immune system cells have been killed by the chemotherapy
- Patient can have an infection (fever only) without shock
- Shock (emergency) occurs when the patient has both a fever and neutropenia
- Patient will either have a very high fever or a very low temperature
- Bacteria is now in the blood stream which can cause the patient to die if not treated early enough

SIGNS AND SYMPTOMS

EARLY RECOGNITION AND IMMEDIATE TREATMENT OF SEPSIS OR SEPTIC SHOCK ARE ESSENTIAL TO A PATIENT'S SURVIVAL!!! YOUR ASSESSMENT IS CRUCIAL!!!

- FEVER or low temperature
- High heart rate and respiratory rate
- LOW blood pressure (approximate examples: 70/30's, 60/20's, or lower)
- Peripheral vasodilation (BOUNDING pulses)
- Mottled hands and/or feet – Capillary refill time greater than 4 seconds



- Reduced mental alertness (hard to wake the patient up)

IMMEDIATE NURSING ACTIONS

- 0.9% NaCl FLUID BOLUS is the MOST IMPORTANT intervention for a patient suffering septic shock
  - 20cc/kg bolus over 5-20 minutes
  - 60cc/kg or greater during the first hour is often needed to stabilize the patient
- Start antibiotics immediately after fluid bolus (usually ceftriaxone or ceftazadime are given first because they cover most bacteria)
- Infection is worse and patient condition is deteriorating if: it is very difficult to wake the patient up, breathing becomes either very fast or very slow, skin is cold and clammy to touch, if you can't feel a pulse anymore or if the pulse is very weak, if the patient stops voiding urine

TUMOR LYSIS SYNDROME

TLS

Hyper means high, above normal (hyperkalemia = higher than normal levels of potassium in the blood)

Hypo means low, below normal (hypocalcemia = lower than normal calcium in the blood)

Symbols:
> means greater than
< means less than

DEFINITION

Tumor Lysis is defined as severe metabolic abnormalities that include hyperuricemia (uric acid >8 mg/dL), hyperphosphatemia (phosphorous >10 mg/dL), hypocalcemia (calcium <8 mg/dL), hyperkalemia (potassium >6 mEq/L), elevated BUN (Blood Urea Nitrogen) and elevated creatinine levels. These abnormalities are a direct result of the death and degradation of tumor cells and the release of their components into the body's circulation or blood stream. These abnormalities may lead to acute renal failure and potentially life threatening heart dysfunction. ACUTE TUMOR LYSIS SYNDROME IS AN ONCOLOGIC EMERGENCY.

Table 1: Risk of TLS Based on Patient Characteristics

- High tumor burden (defined as tumor large in size, LDH >1,500 IU/L, WBC >25,000/mm^3)
- Extensive bone-marrow involvement
- Elevated pretreatment uric-acid levels
- Tumor that is highly sensitive to treatment
- Dehydration
- Decreased urine output
- Acidic urine
- Pre-existing renal dysfunction
- Tumor involvement of the kidney and/or renal vasculature
- Advanced age

TLS: tumor lysis syndrome; LDH: lactic dehydrogenase; WBC: white blood cell.
Source: References 1, 4, 6.

Table 2: Risk of TLS According to Tumor Type

Degree of Risk	Tumor Type
High	Burkitt's lymphoma High-grade non-Hodgkin's lymphoma Lymphoblastic lymphoma T-cell acute leukemia Other acute leukemias
Moderate	Low-grade lymphoma treated with chemotherapy/radiation/corticosteroids Multiple myeloma Breast carcinoma treated with chemotherapy/hormonal therapy Small-cell lung carcinoma Germ-cell tumors (seminoma, ovarian)
Low	Low-grade lymphoma treated with interferon Merkel's cell carcinoma Medulloblastoma Adenocarcinoma of the gastrointestinal tract

TLS: tumor lysis syndrome.
Source: Reference 5.

http://www.uspharmacist.com/content/s/35/c/10134/

PRESENTATION

- TLS occurs quickly and often happens 6-48 hours after initial treatment is begun; it can, though, occur before treatment starts or up to 7 days after treatment is begun.
- Early symptoms may include weakness, muscle cramps, abdominal pain, nausea, vomiting, diarrhea and lethargy and decreased urine output.
- Late symptoms may include hypotension, edema, flank pain, blood in urine, convulsions, slow heart rate, and paralysis.
- If untreated, symptoms will lead to seizures, renal failure, and cardiac arrest.

SPECIAL NURSING CONSIDERATIONS FOR CARING FOR A CHILD WITH TUMOR LYSIS

- Prevention is crucial
- Identify Patients with Risk Factors: Acute Lymphoblastic Leukemia (ALL) and Non-Hodgkin's Lymphoma (NHL) at high risk
- Standard Prevention Protocol:
  - Allopurinol (This is usually given orally; it can be given IV. Allopurinol is ideally prescribed 2-4 days prior to beginning chemotherapy)
  - Hydration (and forced diuresis with furosemide or Mannitol if necessary)
  - Urinary Alkalinization (by adding sodium bicarbonate to IV fluids)
- Early Recognition is also VERY IMPORTANT
- Pay close attention to fluid balance; record intake and output every 4 hours
- Maintain urine output greater than 3-5cc/kg/hour; Administer IV hydration as ordered – not less than 3l/m2/day
- Draw blood samples as ordered; Monitor electrolyte levels; Treat imbalances as appropriate.

HEMORRHAGIC CYSTITIS

DEFINITION OF SPECIFIC TOXICITY

Hemorrhagic cystitis is a diffuse inflammation of the urinary bladder that is evidenced by blood in urine, hematuria (microscopic or macroscopic). Patients frequently experience dysuria (painful urination) and bladder spasms. Severe cases may result in hemorrhage.

COMMON CAUSATIVE AGENTS

- Cyclophosphamide
- Ifosfamide

Hematuria specimens will show levels of red blood cells ,white blood cells, epithelial cells, casts , bacteria, yeast, crystals, pH, glucose, ketones and more.

PREVENTIVE MEASURES

Ensure adequate hydration prior to, during and following the administration of cyclophosphamide and ifosfamide

- IV fluid administration is typically given pre and post administration of these drugs
- Pre-hydration rate is 150 m/m^2 /hr
- Chemotherapy may be initiated when urine output is double the body weight (ml per kg of body weight) in one void or in any one hour after the start of hydration
- After chemotherapy, resume hydration at a rate of 2.5 times maintenance

If mesna has been administered (all patients receiving ifosfamide and those receiving more than 1200mg/m2 cyclophosphamide), continue hydration until the patient is tolerating oral fluids, and oral intake and urine output are normal

- Uroprotection with mesna must be used for patients receiving ifosfamide
- Uroprotection with mesna may be used in patients receiving cyclophosphamide

GENERAL CONSIDERATIONS

- The daily dose of mesna is typically administered in dosages of 20% of the dose of cyclophosphamide or ifosfamide.
- It is administered in four doses
- The first dose may be administered in the same IV solution as cyclophosphamide and ifosfamide and it is usually administered at 3, 6 and 9 hours post the infusion of cyclophosphamide or ifosfamide.

ASSESSMENT

- Perform a complete urinalysis if hematuria develops
- Perform a physical examination if symptoms of dysuria develop
- Rule out possible infectious causes whenever possible

MONITORING

- Ideally, each void or at least one void per 8 hours should be checked for the microscopic presence of red blood cells with the use of urinary dipsticks
- Daily urinalysis including microscopy for detection of the presence of red blood cells
- Visual inspection of urine output for color
- History and physical examination of the patient

HEMORRHAGIC CYSTITIS (continued)

MANAGEMENT

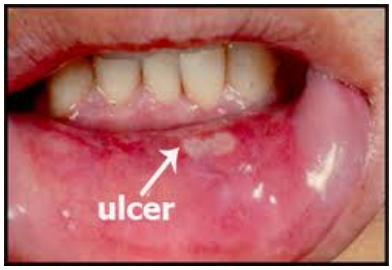
- Microscopic hematuria (any degree), administer mesna with subsequent cyclophosphamide (in patients who have not previously received mesna) in four daily doses at 80% of the total daily dose of cyclophosphamide
- Macroscopic hematuria (gross hematuria) which occurs in patients receiving mesna with either cyclophosphamide or ifosfamide, mesna should be given with the subsequent dose of the chemotherapeutic agent and then given as a continuous infusion and continued for at least 47 hours following the administration of the chemotherapy. If hematuria persists, mesna may be administered at double the dose in the continuous infusion. Adequate IV hydration rates must be maintained
- Symptomatic management for dysuria and/or bladder spasms may be required

MOUTH ULCERS

Mucositis / Stomatitis

DEFINITION OF SPECIFIC TOXICITY

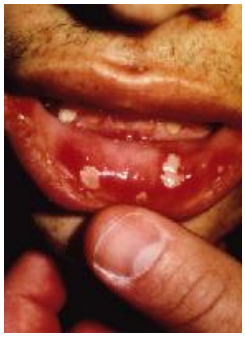
- Mucous membrane lining contains rapidly dividing cells and, therefore, sensitive to chemotherapy
- Presentation: dry, cracked lips; red ulcerated mucous membranes; pain with swallowing; reluctance or inability to eat or drink; saliva - increased in early stages, decreased and thick in advanced stages



COMMON CAUSATIVE AGENTS

- Methotrexate, doxorubicin
- 5FU, daunorubicin, Idarubicin, dactinomycin, cytarabine
- high-dose etopodise, high-dose cyclophosphamide, irinotecan

OTHER POSSIBLE, NON-CHEMO RELATED CAUSES TO CONSIDER



- Poor oral hygiene
- Previous viral infection requiring specific therapy (e.g. HSV)

ASSESSMENT

- Assess oral cavity and nutritional and hydration status
- Pain assessment

MONITORING

- Frequent oral cavity examination, daily weight, nutritional/hydration status
- Pain assessment

PREVENTIVE MEASURES

- Dental evaluation prior to therapy
- Good oral hygiene
    - Mouth rinse with a bland mouthwash that contains no alcohol or a bicarbonate mouth rinse or plain saline.
    - Rinse the teeth and mouth after eating
    - Brush teeth with a soft toothbrush and floss teeth unless platelet count is <50,000 or the ANC is <500

MANAGEMENT

Symptom management

- Oral care as per preventive measures
- Analgesic use
    - Topical agents (e.g. lidocaine)
    - Pain management using opioid medications
- Use of cooling agents (e.g. ice chips, popsicles)

- Depending on severity of mucositis and resultant nutritional support requirements, consider nasogastric feeds if necessary

TYPHLITIS / NEUTROPENIC ENTEROCOLITIS

DEFINITION

- Typhlitis is a necrotizing inflammation of the cecum, sometimes extending into the ileum and ascending colon
- One of the most serious complications of aggressive chemotherapy
- More prevalent when patient is severely neutropenic

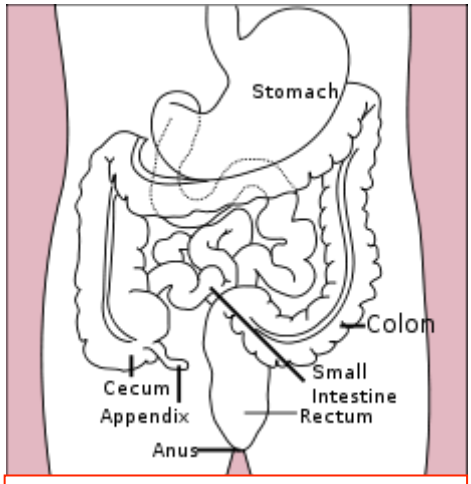


Image Reproduced from http://en.wikipedia.org/wiki/Caecitis

SYMPTOMS

- Fever
- Abdominal pain (usually right lower quadrant)
- Nausea, vomiting
- Diarrhea
- Abdominal distension

SIGNS

- Change in vital signs
- Tachycardia (increase in heart rate)
- Hypotension (low Blood Pressure)
- Abdominal distension
- Rebound tenderness
- Blood in stools

DIAGNOSIS

Xray of the abdomen

- Obstruction
- Pneumatosis
- Free air

TREATMENT

- Conservative medical management
  - Broad spectrum antibiotics with anaerobic coverage
  - Gut rest
  - Supportive care – fluids, blood products
- Surgery only if clinical deterioration in spite of aggressive medical management
- Mortality could be 50-100%

GLOSSARY OF TERMINOLOGY

Adapted from http://www.chemotherapy.com/glossary/terms.html and http://www.chemocare.com/whatis/important_chemotherapy_terms.asp.

ANC (absolute neutrophil count)—ANC refers to the percentage of the total white blood cell count that is made up of cells called neutrophils. Neutrophils are particularly important because they defend our bodies against infection.

AML (acute myeloid leukemia)—AML is a disease in which the bone marrow produces white blood cells that cannot carry out normal function. Signs of the disease include bleeding gums, anemia, fatigue, fever, bone pain, and repeated infections.

Adjuvant chemotherapy—Adjuvant chemotherapy is chemotherapy given after surgery or radiation therapy when there is no visible cancer but there is a risk that there are still cancer cells left in the body.

Alopecia—Alopecia is hair loss. Chemotherapy and sometimes radiation may make patients lose some or all of their hair during treatments. The most common area involved is the head, although other body hair can also be affected.

Analgesic—A medication that relieves pain.

Anemia—Anemia is a lower-than-normal number of red cells in the blood. Red blood cells are important because they carry oxygen from the lungs to all other cells in the body. Shortness of breath, fatigue, and weakness are common signs of anemia.

Antibiotic—An antibiotic is a medication used to fight germs or bacteria that cause infection. Chemotherapy can make patients more at risk for infection. Antibiotics are given to treat an infection.

Antiemetic—An antiemetic is a medication used to stop or help prevent nausea and vomiting, common side effects of some chemotherapy.

Benign—Not cancerous. Benign tumors may also be referred to as nonmalignant.

Biopsy—Removal of a tissue sample from the body to see if the cells are cancerous. A doctor examines the cells under a microscope, comparing them to normal cells. Techniques to remove cells include:

- **Fine needle aspiration (FNA) biopsies** use a needle attached to a syringe to withdraw a small amount of tissue from a tumor. When a slightly larger needle is used, it is called a **needle core biopsy**. Sometimes doctors use an ultrasound or a computed axial tomography, or CT, scan to view the tumor and assist them with needle placement.
- During an **excisional biopsy**, a surgeon removes an entire tumor. During an **incisional biopsy**, only a small amount of tumor is removed. Both of these procedures involve a surgeon cutting through the skin. Sometimes the surgery requires general anesthesia, and sometimes it can be done by simply numbing the area to be cut (local anesthesia).

Cancer—A group of diseases where normal cells change into abnormal cells that grow out of control, invade surrounding tissues and organs, and may spread to distant sites in the body (metastases)

Carcinogen—A carcinogen is anything that causes cancer. Carcinogens can be physical (eg, UV light), chemical (eg, cigarette smoke), or viral, but many are not known.

Carcinoma—Cancer that begins in the skin or in tissues lining or covering internal organs.

Chemotherapy—The use of drugs to destroy cancer cells. A person on chemotherapy may take one drug or a combination of drugs. Most often these drugs are given by vein using intravenous infusion. Some can be taken by mouth or given in a shot.

Combination chemotherapy—Using more than one anticancer medication together, with the goal of destroying more cancer cells.

CBC (complete blood count)—The CBC is a test that determines the number of red blood cells, white blood cells, and platelets in the blood.

CNS (central nervous system)—The brain and spinal cord.

Complete remission—Complete remission, also known as a clinical complete remission, is when physician can no longer see the tumor by simple examination, chest x-rays, and/or blood tests. A partial remission is where some tumor can still be detected.

Consolidation chemotherapy - Chemotherapy given once a remission is achieved. The goal of this therapy is to sustain a remission. Consolidation chemotherapy may also be called intensification therapy. This term is commonly used in the treatment of acute leukemias.

Constipation—Constipation is difficulty passing stool. It can also refer to a decrease in the normal frequency of bowel movements. It may be accompanied by gas, pain, or pressure in the abdomen.

Cycle—Chemotherapy can be given in a variety of time arrangements, such as daily, weekly, or monthly. Chemotherapy is generally given in cycles. A cycle can last 1 or more days but usually lasts 2, 3, or 4 weeks.

Dehydration—A condition caused by the loss of too much water from the body. Causes include severe diarrhea or vomiting.

Dextrose Fluids (D5W)- Dextrose 5% in Water. It is used to supply water and calories to the body. It is also used as a mixing solution (diluent) for other IV medications. Dextrose is a natural sugar found in the body and serves as a major energy source. When used as an energy source, dextrose allows the body to preserve its muscle mass.

Diagnosis—Identification of a condition or disease based on the signs and symptoms, laboratory tests, procedures, history, and physical examination of the patient.

Diarrhea—Bowel movements that occur more frequently and are more liquid in consistency than normal. Chemotherapy, medication, radiation, and infection may cause diarrhea. Diarrhea can also be caused by medications given to prevent nausea or by antibiotics given to treat or prevent infection.

Electrolyte imbalance—Having too many or too few electrolytes in your body. Electrolytes are electronically charged substances that help move nutrients into and waste out of your body and help keep your organs functioning properly. Examples of electrolyte imbalance are hyponatremia or hypokalemia.

Electrolytes—Substances that break down in the body into salts that help move nutrients into cells and help flush waste out of cells. Electrolytes also help keep your heart, nerves, muscles, and brain functioning properly.

Fatigue—Fatigue means feeling tired, weak, sleepy, forgetful, or worn out, and having no energy to go about your daily routine. Fatigue is commonly caused by cancer treatments, but can also result from the disease itself. Fatigue is also often present in patients with anemia.

Febrile neutropenia—Having a fever and a low white blood cell count (neutropenia). Having a fever during neutropenia is often a sign of infection.

First line chemotherapy - Chemotherapy that has, through research studies and clinical trials, been determined to have the best probability of treating a given cancer. This may also be called standard therapy.

Grade—Grade is the measurement of a cancer, reflecting how abnormal the cells look under a microscope. There are several grading systems for cancer, but all divide cancers into those with:

- Least abnormality (grade 1 or well differentiated)
- Intermediate features (grade 2 or moderately differentiated)
- Greatest abnormality (grade 3 or 4 or poorly differentiated)

A specialist called a **pathologist** performs the grading by examining the biopsy specimen. Knowing the grade is important because higher-grade cancers tend to grow and spread more quickly and have a worse prognosis. A cancer's nuclear grade is based on features of the central part of its cells, the nucleus. The histologic grade refers to how much the tumor cells resemble normal cells of the same type of tissue.

Hb (hemoglobin)—The part of the red blood cell that carries oxygen from the lungs to other organs in the body, such as the brain and the heart. A person with a low hemoglobin level may have anemia.

Hct (hematocrit)—A blood test that measures the number of red blood cells in the bloodstream. The lower the hematocrit, the lower the number of red blood cells in the blood. A person with a low hematocrit may have anemia.

Hematopoietic stem cell—Specialized cell in the bone marrow that produces white and red blood cells and platelets.

Hypokalemia—Low potassium levels in the blood.

Hyponatremia—Low sodium levels in the body.

Immune system—The body's defense system against bacterial, viral, and fungal infections. The immune system includes white blood cells and protective barriers such as the skin and mucous membranes. The principal organs of the immune system are the bone marrow, spleen, and lymph system.

Induction chemotherapy - Chemotherapy given to induce a remission. This term is commonly used in the treatment of acute leukemias.

Infection—An invasion of microorganisms that have the ability to multiply and produce disease.

Inflammation—Redness, swelling, pain, and/or a feeling of heat in an area of the body, often due to infection, irritation, or injury.

Infusion—A process of delivering medications, fluids, or blood products into the body through the bloodstream. A needle is used to gain access through a vein, and a catheter with tubing is used to deliver the fluid.

Intramuscular (IM)- into a muscle. An intramuscular medication is given into the body by a needle into a muscle.

Intrathecal (IT)- into the fluid surrounding the brain and spinal cord (cerebrospinal fluid).

Intravenous (IV)—Into a vein. An intravenous medication is delivered into the body through a vein.

Jaundice—A yellowing of the skin caused by abnormal liver function.

Leukemia—Cancer that begins in the cells of blood-forming tissue (eg, bone marrow).

Local therapy—Cancer treatment that only affects a tumor and the area close to it.

Lymph nodes—Lymph nodes are small, oval glands found throughout the body. They act as filters and fight infection. Cancer cells often spread to other parts of the body through the lymphatic system.

Lymphoma—Cancer that begins in cells of the immune system.

Maintenance chemotherapy - Chemotherapy given in lower doses to assist in prolonging a remission. Maintenance chemotherapy is used only for certain types of cancer, most commonly acute lymphocytic leukemias and acute promyelocytic leukemias.

Malignant—Malignant means that a tissue has cancer cells in it that come from a different site in the body; it also refers to a cancerous disease.

Malnutrition—A condition caused by not getting enough calories or nutrients such as vitamins and minerals. Causes of malnutrition include not getting enough nutrients in your diet or not being able to absorb nutrients, all of which can result from cancer or some cancer treatments.

Metastasis—The spread of cancer from one part of the body to another.

Modality—A method of treatment. A multi-modality treatment regimen involves several types of treatment, such as chemotherapy, radiation, and surgery.

Mucositis—Inflammation of the lining of the gastrointestinal tract. **Oral mucositis** refers to inflammation of the lining of the mouth. Mucositis may involve sores, swelling, pain, and redness.

Myelosuppression—Myelosuppression occurs when the bone marrow slows production of blood cells. This results in fewer red blood cells, white blood cells, or platelets available to perform their normal functions in the body. Chemotherapy can cause decreased bone marrow function. Most often, myelosuppression refers to the loss of white blood cells.

Nausea—Feeling queasy or sick to your stomach.

Neoadjuvant chemotherapy—Chemotherapy given before surgery in order to shrink the tumor.

Neutropenia—Neutropenia occurs if there is a lower-than-normal number of neutrophils (infection-fighting white blood cells) in the blood. It is a common side effect of chemotherapy treatment. Neutrophils fight infection, so a person with a low neutrophil count will be more at risk for developing infection. Doctors check the number of neutrophils when they measure the white blood cell count; the result is often referred to as the ANC, or absolute neutrophil count.

Neutrophil—The most common type of white blood cell. Neutrophils help the body fight infection. Since the most common type of white blood cell is the neutrophil, a low white blood cell count usually indicates that the neutrophil count is low. It is easier to get an infection and harder to recover from an infection when the number of neutrophils in the bloodstream is low.

Normal Saline (NS) - 0.9% Sodium Chloride Injection contains 9 g/L Sodium Chloride (NaCl)

NSAID (nonsteroidal anti-inflammatory drug)—Medications, usually available without a prescription, that decrease inflammation (swelling, fever) and reduce pain.

Oncologist—A physician who specializes in the treatment of cancer.

Opioid—A substance used to treat moderate to severe pain that binds to opioid receptors in the central nervous system.

Oral (PO) —When medication is given by mouth.

Palliative care—Palliative care focuses on controlling symptoms and improving quality of life for patients.

Partial remission—Partial remission is a significant decrease in the number of cancer cells, but not their complete disappearance, in response to the cancer therapy. This is in contrast to a clinical complete remission where the tumor cells are no longer observable by, for example simple examination, x-ray and/or blood tests.

Pathology—The study of the causes and characteristics of disease.

Peripheral neuropathy—A possible side effect of some chemotherapy, characterized by numbness, tingling, or burning in the hands and feet.

Platelets—One of the three types of cells made in the bone marrow. The main function of platelets is to aid in clotting the blood following an injury.

Prognosis—A prediction of the likely outcome of a disease based on the current health of the patient and the usual course of the disease.

Proliferate—To reproduce through cell division.

Radiation therapy—A cancer treatment that involves the use of x-rays, gamma rays, and other types of radiation to kill cancer cells, either by directly damaging their DNA or inhibiting their ability to proliferate.

Rectal—A route of administration of therapy where the medication is given through the rectum.

Red blood cell—Red blood cells are made in the bone marrow and released into the blood. They circulate in the blood and carry oxygen and carbon dioxide to and from every cell in the body.

Regimen—A plan of treatment, including doses, scheduling, and duration of treatment.

Remission—The disappearance of a cancer, as determined by clinical evaluation, resolution of symptoms, or both. Complete remission is the disappearance of all signs of cancer after treatment. Partial remission is a notable decrease in cancer cells, but not their complete disappearance, in response to therapy.

Risk factor—Anything that increases the chance of getting a certain disease, such as cancer. Some risk factors can be controlled, such as smoking. Other risk factors, such as age and race, cannot be controlled.

Sarcoma—Cancer of the bone, cartilage, fat, muscle, blood vessels, or other connective or supportive tissue.

Second line chemotherapy - Chemotherapy that is given if a disease has not responded or reoccurred after first line chemotherapy. Second line chemotherapy has, through research studies and clinical trials, been determined to be effective in treating a given cancer that has not responded or reoccurred after standard chemotherapy. In some cases, this may also be referred to as salvage therapy.

Side effect—A change in a person's condition caused by taking a drug, medical device, or other treatment. For example, common side effects of chemotherapy include fatigue, nausea, vomiting, and loss of appetite. Acute side effects are short-term side effects that often go away soon after treatment stops. Chronic side effects are long term.

Single agent—Single-agent therapy refers to the use of one chemotherapeutic medication for the treatment of cancer.

Stage—Staging is a method of determining the extent of the cancer, or how far the disease has spread. The stage is determined after performing a series of diagnostic tests, which may include x-rays, CT/CAT scans, and sometimes surgery. Knowing the stage of the cancer will help your doctor decide the best treatment course.

Subcutaneous (SQ) - into the subcutis, the layer of skin directly below the dermis and epidermis, collectively referred to as the cutis. Subcutaneous injections are highly effective in administering vaccines and such medications as insulin and morphine.

Symptom—A sign or indicator of a disease or illness.

Systemic therapy—Cancer treatment that affects cells throughout the body. Chemotherapy is a systemic treatment.

Sterile Water (SW) or (SWFI) is Sterile Water for Injection- A vial of sterile water that is used to mix with some medications that are powder in order to make a solution.

Thrombocytes—A piece of cell formed from the bone marrow that aids in wound healing and blood clotting. Also called platelets.

Thrombocytopenia—A condition resulting from an abnormally low number of platelets (thrombocytes) circulating in the blood. Bleeding and/or bruising may occur if the platelet count is especially low (less than 20,000/mL).

Topical—A route of administration of therapy where medication is absorbed through the skin.

Transfusion—An intravenous infusion of blood or blood components.

Tumor—A collection of cells that appears as a lump, mass, or swelling.

Tumor markers—Substances in the blood or urine that are associated with particular kinds of cancer. These chemicals can be measured to help doctors diagnose cancer and evaluate the effectiveness of a cancer treatment. A rise in the level of a marker could mean the cancer is growing; a drop in the level could indicate the treatment regimen is effective.

Ulceration—A break on the skin or on the surface of an organ. Ulcers are sometimes associated with cancer.

White blood cell—A white blood cell is one of the three main types of blood cells. White blood cells are responsible for fighting infection. There are several kinds of white blood cells, including monocytes, lymphocytes, neutrophils, eosinophils, and basophils.

X-ray—A test used for diagnosis and assessment. During an x-ray, a small amount of radiation passes through the body and leaves an image of the shape of the internal organs on film.

* All expiration dates apply to dilutions unless otherwise specified.

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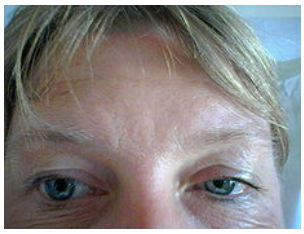
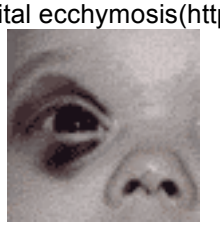
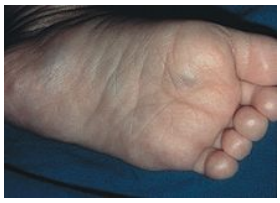
APPENDIX A - Neuroblastoma

Neuroblastoma

- Horner's syndrome (http://en.wikipedia.org/wiki/Horner's_syndrome)
  - **"Horner's syndrome** (also **Horner syndrome**, **Bernard-Horner syndrome**, **Claude Bernard-Horner syndrome** or as **oculosympathetic palsy**) is the combination of drooping of the eyelid (ptosis) and constriction of the pupil (miosis), sometimes accompanied by decreased sweating (anhidrosis) of the face on the same side; redness of the conjunctiva of the eye is often also present. Apparent enophthalmos is also a frequent symptom. It indicates a problem with the sympathetic nervous system, a part of the autonomic nervous system. Medical imaging and response to particular eye drops may be required to identify the location of the problem and the underlying cause.[1]"
  - 
- Periorbital ecchymosis(http://www.medscape.com/viewarticle/405712_2)
  - 
- Pepper Syndrome (http://www.medilexicon.com/medicaldictionary.php?t=88677)
  - 1. obsolete eponym for neuroblastoma of the adrenal gland with metastases in the liver; formerly believed to occur more frequently when the primary tumor was in the right adrenal, whereas tumors of the left adrenal tended to metastasize to the skull (Hutchison syndrome).
- Blueberry Muffin Syndrome (http://en.wikipedia.org/wiki/Blueberry_muffin_baby)
  - **Blueberry muffin baby** is the characteristic distributed purpura occurring as a result of extramedullary hematopoiesis found in infants.[1][2]:826 The purpura is often generalized but favors the trunk, head, and neck.[3] It derives its name from the superficial similarity to a blueberry muffin…It was originally considered characteristic of rubella, but is now considered to be potentially associated with many other conditions,[4] such as cytomegalovirus[5] and metastatic neuroblastoma.
  - 
- **Neuroblastoma 4S** (http://www.cancer.gov/cancertopics/pdq/treatment/neuroblastoma/Patient/page2#Keypoint14)
  - In stage 4S:
    - the child is younger than 12 months; and

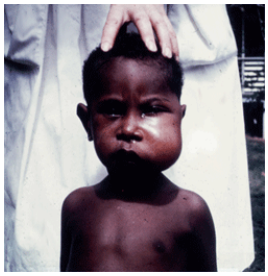
        - the cancer has spread to the skin, liver, and/or bone marrow; and
        - the tumor is in only one area and all of the tumor that can be seen *may* be completely removed during surgery; and/or
        - cancer cells may be found in the lymph nodes near the tumor.
    - There is no standard treatment for stage 4S neuroblastoma but treatment options include the following:
        - Watchful waiting with supportive care for patients who do not have symptoms.
        - Chemotherapy and/or radiation therapy for patients who have symptoms.
        - (http://www.cancer.gov/cancertopics/pdq/treatment/neuroblastoma/Patient/page5#Section_253)

APPENDIX B - BURKITT LYMPHOMA/MALARIA

“5.10.1. Malaria and Burkitt lymphoma

(http://www.impact-malaria.com/web/malaria_training/immunopathology_burkitt-lymphoma_)

Burkitt's lymphoma is the commonest childhood cancer in Africa. The lymphoma is sporadic worldwide and only occurs at high in areas where P. falciparum is highly prevalent, hence the suggestion that the two are linked. This concept is reinforced by the observation that endemic Burkitt's lymphoma disappears from areas where malaria has been controlled.

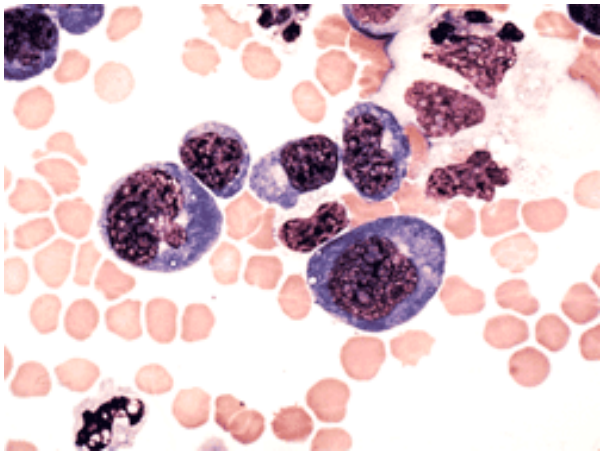


Credits: Herbert Gilles

Burkitt lymphoma in an African child

Mechanism

The basic molecular event in Burkitt's lymphoma is **the translocation of an oncogene** involved in the regulation of the cell cycle (c-myc) from its normal position on chromosome 8 to various other chromosomes, always in close proximity to the regions that regulate the expression of immunoglobulin genes. It has been suggested that polyclonal activation of B cells may de-regulate the oncogene and allow its translocation to take place.



Monomorphic lymphoid proliferation with monocytic infiltration is the characteristic histological picture of Burkitt's lymphoma.

The effects of malaria on host immunity, which include a mixture of intense proliferation of lymphocytes stimulated by malarial antigens and an immunosuppression of mechanisms controlling polyclonal proliferation, creates a suitable environment for the proliferation of some viruses. ***This is true for the Epstein-Barr virus (EBV)***. There is no direct evidence that EBV is the causative agent of Burkitt's lymphoma, but epidemiological studies indicate a strong relationship. ***In areas of co-endemicity, malaria is believed to be a major provoking factor in the evolution of EBV infection towards the malignant Burkitt's lymphoma.***

Relation between the three diseases

A multistep scenario is proposed to explain the development of the lymphoma:

- chronic malaria results in the polyclonal activation of B cells
- polyclonal activation increases the chances of c-myc translocation
- EBV infection of B-cells where a translocation has taken place induces autonomous growth of these cells
- malaria-induced immunosuppression decreases the efficiency of anti-EBV immunity (for example, reduced EBV-specific cytotoxic T-cells in malaria)"

APPENDIX C - T-CELL LYMPHOMA

Overview

(http://www.lymphoma.org/site/pp.asp?c=bkLTKaOQLmK8E&b=6300161)

Lymphoma is the most common blood cancer. The two main forms of lymphoma are Hodgkin lymphoma and non-Hodgkin lymphoma (NHL). Lymphoma occurs when cells of the immune system called lymphocytes, a type of white blood cell, grow and multiply uncontrollably. Cancerous lymphocytes can travel to many parts of the body, including the lymph nodes, spleen, bone marrow, blood, or other organs, and form a mass called a tumor. The body has two main types of lymphocytes that can develop into lymphomas: B-lymphocytes (B-cells) and T-lymphocytes (T-cells). T-cell lymphomas account for approximately 15 percent of all NHLs in the United States. A similar lymphocyte called a natural killer (NK) cell shares many features with T-cells. When NK cells become cancerous, the cancer is called NK or NK/T-cell lymphoma and is generally grouped with other T-cell lymphomas. There are many different forms of T-cell lymphomas, some of which are extremely rare. T-cell lymphomas can be aggressive (fast-growing) or indolent (slow-growing). Lymphomas are often, but not always, named from a description of the normal cell that leads to cancer. The general term peripheral T-cell lymphoma (PTCL) refers to the entire group of mature or "post-thymic" T-cell lymphomas (arise from mature T-cells), which distinguishes them from the immature T-cell lymphomas such as acute lymphocytic leukemia (ALL) or lymphoblastic lymphoma. Under this broad meaning, almost all types of T-cell lymphoma fall under the category of PTCL.

APPENDIX D - LYMPHOMA/HIV

Non-Hodgkin's lymphoma

(http://mobile.aidsmap.com/Non-Hodgkins-lymphoma/page/1044727)

Michael Carter

Published: 06 June 2012

"Non-Hodgkin's lymphoma is an AIDS-defining cancer. A lymphoma is the name given to a tumour (or growth) of lymphocytes (white blood cells). Non-Hodgkin's lymphoma (NHL) is caused by the unregulated production of B-cells, and is sometimes called B-cell lymphoma.

B-cells are one of the two main classes of lymphocytes, (the other being the T-cells). They are produced in the bone marrow and spleen and are involved in the production of antibodies. In HIV infection, B-cells typically become 'over-active'. People who are infected with Epstein-Barr virus (which also causes glandular fever), may develop a generalised increase in B-cell reproduction. In some people, particularly if the immune system is suppressed, the continuous replication of B-cells may cause lymphoma.

NHL may occur in the lymph nodes (glands), spleen, digestive system, liver, kidney or – in a particular form seen in immuno-suppressed people – in the brain, where it often occurs without any further spread and is called primary CNS (central nervous system) lymphoma.

Although lymphomas can occur at any CD4 count, they are more common in people with very low counts. NHL in people with HIV can be more aggressive and respond less well to treatment than in HIV-negative people.

With the advent of modern HIV treatment many AIDS-related illnesses have become less common and NHL is now less commonly seen in people with HIV."

APPENDIX E – INFECTION

Causes of Immunosuppression in child with cancer

(Kline, N. (2007) Prevention and Treatment of Infections. In: Nursing Care of Children and Adolescents with Cancer 3rd. ed. 267-278. Glenview, IL: APON)

"Cancer therapy contributes to many deficiencies in local and systemic host defense (Box 10-1). Lymphocyte function can be impaired by several factors: stem cell defects preventing normal lymphocyte development and causing failure of the immune system; dysfunction of a main lymphoid organ (e.g., the spleen), preventing maturation of stem cells into T or B cells; or disruption of the final stages of B cell maturation. Alterations in the inflammatory response (e.g., chemotactic[1] and phagocytic[2] activities of neutrophils and macrophages) can also impair host resistance.

Children with T cell defects are prone to viral infections (E.g., varicella, herpes simplex, cytomegalovirus) and fungal infections (e.g., *Cryptococcus, Candida spp*.), as well as *Pneumocystis carinii*.

The duration of neutropenia is often indicative of the risk of infection. Less than 30% of patients with neutropenia with duration less than one week have evidence of infection, compared with nearly 100% of patients when neutropenia lasts longer than 1 week.

Management of the child with a fever and neutropenia differs markedly from that of other patients with fever. **Because of the high mortality rate associated with untreated infection, all fevers experienced by children with neutropenia are considered due to a life-threatening infection unless proven otherwise.** The initial clinical signs and symptoms of infection (e.g., erythemia, exudate, swelling, localized adenopathy) in neutropenic patients with either unexplained fever or septicemia can be blunted or absent, as white blood cells are needed for these processes.

At the onset of fever, it is crucial to perform a careful but expeditious evaluation before initiating antibiotic therapy. The lung is the most frequent site of serious infection, followed by soft tissues, mucosa, and the blood. Urinary tract infections are less common in children than in adults, and infections of the central nervous system are unusual.

- Because the classic signs and symptoms of infection often are absent, subtle signs of inflammation must be noticed.
- Detailed questioning about cough, shortness of breath, tachypnea, pain with defecation, skin rashes and lesions, odynophagia (inability to swallow), sore throat, and retrosternal pain is necessary.
- A meticulous physical examination must be done so that any subtle signs of infection or inflammation are not missed.

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[1] Chemotactic factor = Chemoattractant, chemotaxin Immunology Any small molecule that acts as a chemical stimulus along a concentration gradient, attracting macrophages and other cells–eg, to a site of inflammation (http://medical-dictionary.thefreedictionary.com/chemotactic+factor)

[2] Phagocyte = A cell, such as a white blood cell, that engulfs and absorbs waste material, harmful microorganisms, or other foreign bodies in the bloodstream and tissues. (http://www.thefreedictionary.com/phagocytic)

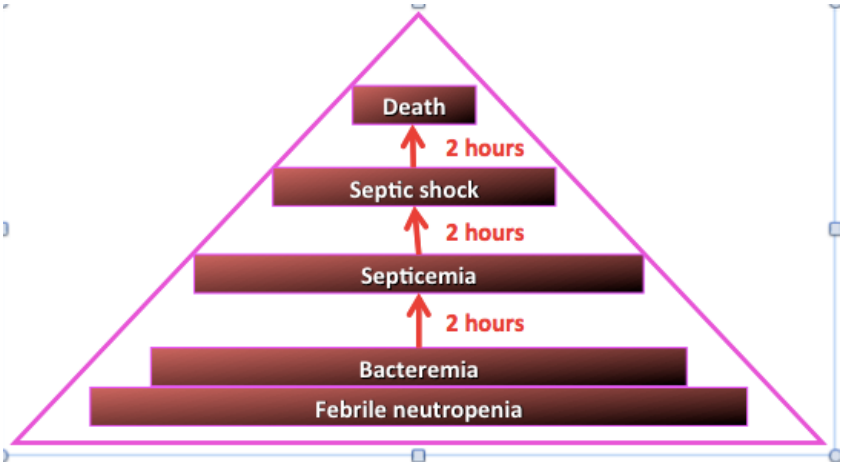
- Blood pressure, heart rate, and respiratory rate are carefully monitored.
- Special attention is given to
  - the oral cavity and posterior oropharynx for ulcers or lesions;
  - fingers and toes for paronychia (infection of the skin around the nail);
  - axillae and groin for rashes or lesions; perianal and vulvar areas for lesions or fissures;
  - and sites of previous procedures such as intravenous or urinary catheters, fingersticks, and bone marrow or other biopsy sites.
- Relatively innocuous-appearing lesions can be the focus for bacteremia or sepsis.
- The nurse must remove all dressings and bandages to examine the skin beneath." *(reformatted from original)*

Table 1.

Bacterial growth and clinical manifestations in neutropenic patients

Time (hours)	Organisms per mL	Clinical Manifestations
0	1	None
0.5	2	None
1.0	4	None
2.0	16	None
4.0	256	None
6.0	4096	Fever
8.0	65,536	Sepsis
10.0	1,048,576	Septic shock
12.0	16,777,216	Death

Table 2. Time sequences



Slides from Scott Howard, M.D. St. Jude Children's Research Hospital with permission

APPENDIX F - WHO MUCOSITIS SCALE