

Glossary of Medical Terms

Α

Absolute Neutrophil Count (ANC): The real number of white blood cells (WBCs) that are <u>neutrophils</u>. Neutrophils are key components in the system of defense against infection. Having few neutrophils present is called <u>neutropenia</u>.

Acyclovir: An anti-viral drug used to treat or prevent cytomegalovirus and herpes simplex infections.

Adenovirus: A set of viruses that induce respiratory tract and eye infections. Gene therapy uses adenoviruses that are modified to carry a particular tumor-fighting gene.

Adrenal glands: Glands located on top of each kidney that secretes several important hormones into the blood. The inner portion of the adrenal gland, the adrenal medulla, stores and releases catecholamines that are measured by testing urine samples.

Alkylating Agents: A class of anticancer drugs that interferes with the cell's DNA and restrains or halts cancer cell growth.

Allogeneic: (Bone Marrow or Peripheral Blood Stem Cell Transplant) a transplant using bone marrow or stem cells from a matched sibling donor infused into recipient after high dose chemotherapy.

Angiogenesis: Blood vessel formation. Angiogenesis involves the growth of blood vessels from surrounding tissue into a solid tumor. **Antiangiogenesis** drugs are drugs that prevent the growth of new blood vessels into a solid tumor.

Anthracyclines: A class of antibiotics used in many induction chemotherapy regimens for high-risk neuroblastoma. It is important to keep track of the cumulative dose of anthracyclines a child receives because these drugs can damage the heart muscle.

Antibiotics: Drugs used to fight bacterial infections.

Antibody: A substance made by the immune system, used to defend the body against bacteria, viruses, toxins or tumors. Neuroblastoma treatment may include the use of man-made monoclonal antibodies (MoAb) in an effort to train the immune system to fight neuroblastoma.

Antiemetics: Drugs used to control nausea and vomiting.

Apheresis: Procedure in which blood is withdrawn from the patient and circulated through a machine that removes specific components, such as stem cells or platelets, and returns the remaining blood components to the patient. High risk neuroblastoma patients have peripheral blood stem cells collected through apheresis for use in stem cell transplant or after certain other types of therapy.

Apoptosis: Programmed cell death.

Ascites: Excess fluid in the abdominal cavity, which causes swelling. Ascites is a possible complication of surgery to remove abdominal tumors and a symptom of VOD.

Ataxia: A problem of muscle coordination causing loss of balance. Ataxia is sometimes seen in neuroblastoma with opsoclonus/myoclonus syndrome (OMS).

Audiogram: A graph showing a person's hearing capacity, based on a set of tests examining perception of distinct sound frequencies.

Autologous bone marrow or peripheral stem cell transplantation (ABMT or auto PBSCT): Transplant in which the patient receives his or her own marrow or stem cells.

В

Benign: Not-malignant.

Beta-glucan: Sugar molecule derived from yeast or barley, used with 3F8 antibody therapy to enhance immune response against neuroblastoma.

Bile: A yellow-green liquid made in the liver whose purpose is to eliminate waste from the liver and break down fats as food is digested.

Bilirubin: The orange-yellow pigment in bile. Jaundice occurs if bilirubin builds up in the blood and skin. Bilirubin can be measured to check for liver disease.

Biological response modifiers: Substances (natural or man-made) that increase or revive healthy immune defenses. Granulocyte Colony Stimulating Factor (GCSF) is often given after chemotherapy to quicken white cell count recovery, and Granulocyte Macrophage Colony Stimulating Factor (GMCSF) is a biological response modifier often used with antibody therapies.

Biopsy: Surgical removal of tissue for examination by a pathologist to determine diagnosis.

Blood type: Identification of a person's blood according to the kinds of proteins present on the surface of the red blood cells.

Bolus: One dose of drug usually injected into a blood vessel over a short time period.

Bone Marrow: The spongy material that fills the inner cavities of the bones and makes all types of blood cells.

Bone Marrow Aspiration: The removal and examination of a small portion of bone marrow cells by suctioning into a syringe.

Bone Marrow Biopsy: The removal and examination of a small portion of bone using a hollow biopsy needle.

Bone Marrow Harvest: A process that collects healthy bone marrow to be stored and used at a later date for a bone marrow transplant.

C

Carboplatin: Platinum compound (<u>DNA alkylating agent)</u> chemotherapy commonly used in frontline therapy for neuroblastoma; less toxic to hearing than cisplatin.

Cardiomyopathy: A disease of the heart muscle. It can be caused by the toxic effects of some anticancer drugs.

Catecholamine: Any of many substances (such as epinephrine, norepinephrine, and dopamine) that function as hormones or neurotransmitters or both. Most neuroblastomas will cause an increase in urinary catecholamines.

Central line (central venous line, CVL): A catheter (tube) that is passed through a vein to end up in the chest portion of the vena cava (the large vein returning blood to the heart) or in the right atrium of the heart.

Central Nervous System (CNS): the largest part of the nervous system, including the brain and the spinal cord.

Chimeric Antibodies (ch 14.18): part-mouse and part-human anti-GD2 antibody.

Cisplatin: Platinum compound (<u>DNA alkylating agent)</u> chemotherapy commonly used in frontline therapy for neuroblastoma; can cause hearing damage.

Complete response (CR): A term for labeling response to treatment, in this case, no detectable disease present.

Computed tomography scanning (CT scanning): A technique for producing cross-sectional images of the body which may show cancer more accurately than other imaging methods in some circumstances. CT passes x-rays through the body at different angles and these images are analyzed by a computer.

Conditioning Regimen: The chemotherapy and/or radiation given transplant patients to destroy remaining cancerous cells and/or create space for healthy new marrow to be transplanted.

Consolidation: A course of treatment with chemotherapy given to the patient while in remission to further reduce the number of cancer cells.

Creatinine: A waste product of protein metabolism that is filtered from the blood by the kidneys and expelled in urine. Creatinine can be measured to assess kidney function.

Cyclophosphamide (brand name Cytoxan): Alkylating chemotherapy in the nitrogen mustard family, commonly used in frontline and relapsed neuroblastoma; can cause bleeding in the bladder, so bladder protectant Mesna is given also.

Cytogenetics: A branch of biology that studies the structure of chromosomes.

Cytomegalovirus (CMV): a herpes virus that occurs in healthy individuals without causing symptoms. In immune compromised individuals, CMV may cause serious illness including retinitis (inflammation of the retina), pneumonia, colitis (inflammation of the large bowel), and encephalitis. Blood products are screened to prevent passing CMV to cancer patients.

Debulking Surgery: Surgically removing as much of the tumor as possible.

Differentiation: The process of maturation of a cell line of cancer cells. When fully differentiated, the cells more closely resemble the normal cells in the tissue of origin. In high-risk neuroblastoma, 13 cis-retinoic acid (Accutane) is used as a differentiation therapy to encourage any remaining undetectable neuroblasts to mature into normal cells.

Diploid DNA: A tumor characteristic in which the basic chromosome number in the neuroblastoma cells is doubled (also referred to as DNA Index = 1.0). This is an unfavorable prognostic factor.

DNA Index: A measurement of the amount of DNA material in neuroblastoma cells. An increase in the number of chromosomes is called hyperdiploid DNA. Diploid is the same as DNA index = 1.0; triploid is DNA Index = 1.5; tetraploid is DNA index = 2.0. Diploid and tetraploid are unfavorable prognostic characteristics, and triploid is favorable.

DNA: Deoxyribonucleic acid; responsible for passing genetic information in nearly all organisms. DNA passes hereditary characteristics and information on cell growth, function, and division. Tumor cells have damaged DNA.

Dopamine: A catecholamine hormone and neurotransmitter that transmits messages in the brain and plays a role in movement. Dopamine is a precurser of adrenaline and noradrenaline.

Doxorubicin (brand name Adriamycin): an <u>anthracycline antibiotic</u> chemotherapy drug that interacts with DNA, commonly used in frontline and relapsed neuroblastoma. Can cause heart damage.

\mathbf{E}

Echocardiogram: A test to image the heart and surrounding tissues that uses high-frequency (ultrasound) sound waves.

Electrocardiogram (EKG or ECG): A record of the electrical impulses that trigger the heartbeat, recorded on a moving strip of paper.

Electrolyte: Minerals, such as sodium potassium, that are found in the blood plasma and must be maintained at certain levels to prevent organ malfunction.

Enteral: General term for intestines.

Epinephrine: Another name for adrenaline. Epinephrine is a hormone produced by the adrenal glands in response to stress, exercise, or fear.

Erythrocyte Sedimentation Rate (ESR): A measure of the time it takes for red blood cells to collect at the bottom of a sample of blood. It is a non-specific sign of inflammation, and can be caused by many diseases such as arthritis or wide-spread cancer. It is also called "sed rate."

Esthesioneuroblastoma: A malignant tumor distinct from neuroblastoma (not a peripheral neuroblastic tumor or pNT) arising from the olfactory epithelium of the superior nasal cavity and cribriform plate. It is rare and its cause is unknown. The type of treatment depends on tumor size and location.

Etoposide (VP-16): An inhibitor of the <u>enzyme topoisomerase II</u>, chemotherapy commonly used in frontline and relapsed neuroblastoma. Can cause leukemia.

F

Febrile: Feverish; with fever.

Ferritin: An iron storage protein that is found especially in the liver and spleen.

<u>Fluorescent</u> <u>in situ</u> <u>hybridization</u> (FISH): A <u>cytogenetic</u> technique which can be used to detect disseminated neuroblastoma cells in blood or bone marrow aspirate and can distinguish between malignant and benign cells. This method localizes the presence or absence of specific <u>DNA sequences</u> on <u>chromosomes</u>. It uses <u>fluorescent probes</u> which bind only to those parts of the chromosome with which they show a high degree of sequence similarity.

G

Ganglion: A <u>tissue mass</u>, which is composed mainly of <u>somata</u> and <u>dendritic structures</u> (parts of nerve cells), which often interconnect with each other to form a complex system of ganglia known as a <u>plexus</u> (network). These structures provide relay points and intermediary connections between different neurological structures in the body, such as the <u>peripheral</u> and <u>central</u> nervous systems.

Ganglioneuroblastoma (GNB): A cancerous growth composed of nerve fibers and mature ganglion cells. It is regarded by many as a differentiated neuroblastoma. Nodular ganglioneuroblastoma has "nodes" of neuroblastoma.

Ganglioneuroma: A neuroma (benign) derived from ganglion cells. Ganglioneuromas are often found in the hands or wrists.

Ganglioside GD2: GD2 is a serum marker found in large amounts on the surface of some neuroblastoma cells. Some institutions are researching the use of antibodies that attack GD2 while limiting damage to healthy cells.

Gastric Tube (G Tube): A tube inserted through a small incision in the abdomen into the <u>stomach</u> and is used for long-term enteral nutrition.

Granulocyte: A type of white blood cell that protects the body against bacterial infections. Patients receiving high dose chemotherapy may receive granulocyte colony stimulating factor (GCSF, Neupogen, or Neulasta) to help the immune system recover more quickly.

Н

Hematocrit: The percentage of total blood volume that consists of red blood cells.

Hematoma: A collection of blood from a broken blood vessel.

Hematopoietic: To make blood, another word for blood-forming stem cells.

Hematuria: Blood in the urine. Gross hematuria means the blood is obvious; microscopic hematuria means the blood is hidden.

Hemoglobin: The pigment in red blood cells that carries oxygen to tissues; hemoglobin bound to oxygen gives blood its red color. A complete blood count (CBC) reports hemoglobin as a measure of red blood cells.

Hemorrhagic cystitis: Bladder ulcers; ifosfamide and cyclophosphamide can cause bleeding in the bladder, so chemo-protectant Mesna is given to protect the bladder.

Hepatomegaly: Enlargement of the liver.

Hickman Catheter: A type of central venous line which consists of a flexible plastic tube inserted into the large vein above the heart, used for administering IV drugs and drawing blood samples.

Hirschsprung's Disease: A congenital condition in which nerve cells do not develop in parts of the intestine, causing the colon to function poorly.

Histology (histopathology): the study of <u>tissue</u> sectioned as a thin slice, using a <u>microtome</u>. It can be described as microscopic <u>anatomy</u>. <u>Histopathology</u> is the microscopic study of diseased tissue, is an important tool of <u>anatomical pathology</u> since accurate diagnosis of <u>cancer</u> and other diseases usually requires histopathological examination of samples. This gives prognostic information about neuroblastomas, and is an important part of risk assessment.

Homovanillic acid (HVA): A dopamine metabolite. HVA is excreted in human urine. High HVA concentrations can be an indicator of active neuroblastoma.

Horner's Syndrome: A complex of abnormal findings marked by sinking in of the eyeball, contraction of the pupil, drooping of the upper eyelid, and vasodilation and anhidrosis of the face. Horner syndrome is caused by injury to the cervical sympathetic nerve fibers on the affected side.

Hot Antibodies (131I-3F8): radioactive iodine connected to 3F8 antibodies that deliver radiation directly to the neuroblastoma, also called radioimmunotherapy.

Human Anti-Mouse Antibodies (HAMA): antibody produced by immune system in response to mouse antibodies 3F8.

Human Leukocyte Antigen (HLA): Genetic information on the surface of white blood cells and platelets. HLA is composed of proteins that play an important role in activating the body's immune system to respond to foreign organisms. HLA typing is done to identify potential donors as a match to provide bone marrow or stem cells to a patient who cannot use their own stem cells for stem cell transplant (allogenic).

Humanized monoclonal antibody (hu 14.18-IL2): this antibody combination is called a fusion protein. The humanized antibody retains only 2% mouse antibody and is fused to interleukin-2. The patient does not form an antibody to this because it is humanized. The antibody delivers the interlekin-2 directly to the neuroblastoma cell, which creates an immune response against the tumor.

I

Ifosfamide: Alkylating chemotherapy in the nitrogen mustard family, less commonly used in frontline and sometimes in relapsed neuroblastoma; can cause bleeding in the bladder, so bladder protectant Mesna is given also.

Immune System: The network of cells and organs that protect the body against infection or disease. Activation of this system against foreign substances is referred to as the immune response. The ability to produce cells that work to combat infection or disease is referred to the immune function.

Immunocompromised: Having a depressed immune system. A person can become immunocompromised because of certain diseases or treatments.

Immunocytology (Immunohystochemistry): Cell surface antigen detection technique using one or more mouse antibodies. This is used to detect very small amounts of neuroblastoma cells in peripheral blood or bone marrow aspirate.

Immunodeficiency: The reduced capacity of the body to combat infection and disease.

Immunoglobulins: Proteins that perform as antibodies.

Immunoscintigraphy: An imaging technique in which antibodies labeled with radioactive substances are administered, and then a picture is taken of areas in the body in places where the antibody localizes.

Immunotherapy: Treatment to activate or return the capacity of the person's immune system to combat infection and disease.

Incidence: The total of new cases of a disease diagnosed annually.

Indolent: A slow growing cancer.

Induction Therapy: Treatment that is used as a first step to shrink a tumor and assess it's response to drugs. Additional therapy is given after induction therapy to destroy remaining cancer.

Infusion: The administration of drugs and other fluids into the blood stream.

Insufion: Trade name of an infusion cannula allowing multiple subcutaneous injections through the same injection port, reducing needle pain that usually comes from frequent daily injection therapy.

Integrative Medicine: Focus to complement mainstream medical care and address the emotional, social, and spiritual needs of patients and families; includes herbal medicine, music, aroma, visual therapy.

Interferons: Substances that can increase the body's normal response to disease (biological response modifiers). Interferons are normally produced by the body, but they can be made in the lab for use in treating cancer. Interferons hinder the division of cancer cells.

Interleukins: Substances that can increase the body's normal response to disease (biological response modifiers) that aid the immune system to combat infection and cancer. Interleukins are normally produced by the body, but they can be made in the lab for use in treating cancer.

International Neuroblastoma Response Criteria (INRC): Definitions of response to NB treatment used in trial results include complete response (CR), very good partial response (VGPR), partial response (PR), mixed response (MR), no response or stable disease (NR or SD), progressive disease (PD).

Intracranial Tumors: Tumors arising in the brain.

Intraoperative Radiation Therapy (IORT): Radiation aimed directly at a tumor during a surgical procedure.

Intraperitoneal: Within the area containing the abdominal organs (the peritoneal cavity).

Intrathecal: The thin space between the lining of the brain and spinal cord. Children with central nervous system (CNS) disease may receive intrathecal therapy using liquid radiation or antibodies administered into this space.

Intravenous (IV): Injection of fluids into a blood vessel.

Iodine-131-Metaiodobenzylguanidine (I-131-MIBG): a radioactive isotope of iodine connected to a compound that is selectively taken up in neuroblastomas and pheochromocytomas.

Irinotecan (CPT-11, Camptosar): A <u>topoisomerase 1 inhibitor</u> chemotherapy drug that is frequently used in relapsed neuroblastoma. It is a semisynthetic analogue of the natural alkaloid camptothecin.

Isotretinoin: A member of the retinoid family of drugs. Also known as 13-*cis*-retinoic acid, or by the trade name Accutane, Amnesteem, Roaccutane, and Claravis.

J

Jaundice: A condition characterized by yellowing of the skin, the whites of the eyes, and a darkening of the urine. Jaundice indicates that the liver is not working properly.

K

Karyorrhexis: The destructive fragmentation of the <u>nucleus</u> of a dying <u>cell</u>. It can occur either as a result of <u>programmed</u> cell death or <u>necrosis</u>.

Kidneys: A pair of organs found in the abdomen. The kidneys remove waste from the blood, which leaves the body as urine.

Killer Cells: A type of white blood cell that attacks tumor cells and other cells that have been invaded by foreign substances.

L

Lactate dehydrogenase (LDH): An enzyme present in most tissue, often used as a marker of tissue breakdown. This can be used as a marker for disease progression in neuroblastoma in some cases.

Laparoscopy: The insertion of a laparoscope (a thin tube with an attached light) through the abdominal wall to view the inside of the abdomen. Laparoscopy is also used to remove tissue samples.

Laparotomy: An incision made in the wall of the abdomen.

Lasix: a loop diuretic that prevents system from absorbing too much salt, allowing the salt to instead be passed in your urine. Lasix uis used to treat fluid retention (edema).

Leukocytes: Cells that help the body combat infections and diseases. Also called white blood cells.

Lugol's (SSKI): Potassium and iodine solution used to protect thyroid from MIBG scans and MIBG therapy

Lumbar Puncture: The penetration of a needle into the lower part of the spinal column to gather cerebrospinal fluid or to give chemotherapy drugs intrathecally. Also called a spinal tap. This procedure is not recommended during tests for diagnosis of suspected neuroblastoma due to possibility of spread of disease to central nervous system.

Lymphatic System: The tissues and organs that produce, store, and carry white blood cells that combat infection and other diseases. This network includes the bone marrow, spleen, thymus, and lymph nodes and a system of thin tubes that carry lymph and white blood cells. These tubes branch, like blood vessels, into all the tissues of the body.

M

Magnetic Resonance Imaging (MRI): A procedure in which a magnet linked to a computer is used to create detailed images of areas inside the body.

Malignant (Malignancy): A cancerous growth with a tendency to invade and destroy nearby tissue and spread to other parts of the body.

Mediastinum: The area between the lungs, which includes the heart, trachea, esophagus, bronchi, and lymph nodes.

Mediport: (brand name) a venous catheter in the chest that has access under the skin.

Megestrol (megace): A hormone that is used to increase appetite in people with cancer.

Melphalan: A chemotherapy drug that belongs to the family of drugs called alkylating agents.

Melphalan: Alkylating chemotherapy in the nitrogen mustard family, commonly used in stem cell transplant regimens for neuroblastoma; can cause severe mucositis.

Meninges: The three membranes that cover and protect the brain and spinal cord.

Mesna: A drug that helps safeguard the kidneys and bladder from the damaging effects of the chemotherapy drugs ifosfamide and cyclophosphamide.

Meta-iodobenzylguanidine (MIBG or mIBG): synthetic analogue of the neurotransmitter norepinephrine; selectively taken up (metabolized) by neuroblastoma cells.

Metastasis: Cancer that has spread from one part of the body to another.

Minimal Residual Disease (MRD): Undetectable cancer cells left behind after treatment that cause relapse. Accutane (cis-retinoic acid) and antibodies are treatments for minimal residual disease.

Mitosis: The process in which a cell duplicates its chromosomes to generate two, identical cells.

Mitosis-karyorrhexis index (MKI): A count of dividing tumor cells for classifying tumor pathology and one factor used for neuroblastoma risk assessment. A high MKI (≥200/5,000 cells) at any age is unfavorable. A low MKI (<100/5,000 cells) for those <60 months of age) is favorable.

Mixed response: Greater than 50% decrease of any lesion with less than 50% decrease in any other as defined by the INRC.

Monoclonal Antibodies (MoAb): Substances made in a laboratory that can locate and attach to cancer cells in the body. Monoclonal antibodies are used to fight cancer by recognizing certain proteins on specific cancer cells. They can be used alone, or used to deliver drugs or radioactive material directly to a tumor. Types of monoclonal antibodies used in neuroblastoma treatment include 3F8, 8H9, hu14.18 and ch14.18.

Monoclonal Antibody 3F8: 3F8 was produced by white blood cells of mice, and it must be carefully prepared for human use. It attaches to GD2, which is a marker on the surface of neuroblastoma cells. 3F8 is part of the standard treatment for high-risk neuroblastoma and is only available at Memorial Sloan-Kettering.

Monoclonal Antibody 8H9: A murine (mouse) IgG1 antibody. The 8H9 antibody is highly reactive with a range of neoplastic tissue, including human brain tumors, childhood sarcomas, and neuroblastomas. The majority of primary brain tumors tested positive with 8H9 antibody. The 8H9 antigen is expressed on cell membranes of a broad spectrum of tumors of neuroectodermal, mesenchymal, and epithelial origin.

Monocyte: A type of white blood cell.

Morphology: The science of the structure and form of organisms.

Mucositis: A complication of cancer treatment that causes the lining of the digestive tract to become inflamed. Mucositis sores can develop in the mouth, throat, stomach or intestinal tract.

Multimodal Treatment: Approach to therapy that uses more than one type of treatment.

MYC-N Amplification: myc myelocytomatosis viral related oncogene, when amplified (more copies) unfavorable prognostic factor for neuroblastoma; same as MYCN **Mycostatin:** A drug that treats fungal infections.

Myelin: The fatty material that covers and protects nerves.

Myeloablative Therapy: Chemotherapy and/or radiation that destroy the blood-producing cells in the bone marrow and requires blood stem cells to recover.

Myelodysplasia: Abnormal bone marrow cells that may lead to a type of leukemia.

Myelofibrosis: A condition in which bone marrow is replaced by fibrous tissue.

Myelosuppressive Therapy: Treatment that inhibits blood cell production.

N

Naso-gastric (NG) Tube: A plastic tube inserted through the <u>nose</u>, past the <u>throat</u>, and down into the <u>stomach</u>, used for feeding and administering drugs.

Neoplasm or Neoplasia: A new growth of benign or malignant tissue.

Nephrectomy: Surgical removal of the kidney.

Neuroblastoma: A malignancy that arises in immature nerve cells, occurring in mostly infants and children.

Neuroectodermal Tumor: A tumor of the central or peripheral nervous systems.

Neuroendocrine: Refers to the nervous system and the endocrine system and their hormones.

Neuropathy: A term used to describe changes in the peripheral nervous system. Some drugs used in treatment of neuroblastoma can cause numbness, tingling, or pain in extremities.

Neurotoxicity: The propensity of some treatments to damage the nervous system.

Neutropenia: An abnormal reduction in the number of infection-fighting white blood cells called neutrophils. Neutropenic patients have no ability to fight infections and generally require hospitalization if they develop a fever.

Neutrophil: A type of white blood cell necessary for fighting infection..

N-MYC Amplification: myc myelocytomatosis viral related oncogene, when amplified (more copies) unfavorable prognostic factor; same as MYCN

No Evidence of Disease (NED): No detectable disease.

No Response: Less than 50% decrease but less than 25% increase in any lesion as defined by the INRC.

Nonsteroidal anti-inflammatory drugs: A group of drugs that decrease swelling, pain, and redness.

Nystatin: A drug that treats fungal infections.

O

Octreotide: A drug similar to the naturally-occurring growth hormone inhibitor somatostatin, used for scans and therapy like MIBG.

Ommaya reservoir: a plastic, dome-shaped device surgically placed under the scalp with thin tubing that passes through an opening in the skull for delivering drugs to the brain and spinal cord.

Oncogene: A gene that normally directs cell growth. If an oncogene is altered, it can encourage or permit the uncontrolled growth of cancer.

Ondansetron (Zofran): A drug that prevents or diminishes nausea and vomiting.

Opsoclonus-Myoclonus Syndrome: a syndrome which includes ataxia and encephalopathy. "Opsoclonus" is an unusual disorder of eye movement in which both eyes dart involuntarily (dancing eyes). "Myoclonus" simply means brief muscle jerks and "ataxia" indicates incoordination..

Ototoxic (ototoxicity): Damaging to hearing. Some chemotherapy and antibiotics can cause hearing damage and are termed ototoxic drugs.

Overall Survival (OS): The percentage of subjects in a study who have survived for a defined period of time. Usually reported as time since diagnosis or treatment. Also called the survival rate. **P**

p53 Gene: A tumor suppressor gene that normally inhibits the growth of tumors that has been found to be altered in many types of cancer.

Palliative Therapy: Treatment given to alleviate symptoms caused by cancer or its treatment with the goal to improving the quality of life.

Palpation: Examination by feeling an area of the body with the fingers to feel the organs or tissues underneath.

Partial Response (PR): The shrinking, but not entire disappearance, of a tumor in response to treatment, defined by the INRC as greater than 50% decrease in measurable disease and 1 or no positive bone marrow site.

Pathologic Fracture: A fracture in a bone to an area that has been weakened by cancer.

Peripheral Blood Stem Cell Transplant (PBSCT): Stem cells collected from the peripheral blood as opposed to directly from bone marrow, and used to "rescue" patient from high-dose chemotherapy and/or radiation.

Peripheral IV line: A short <u>catheter</u> inserted through the skin into a <u>peripheral vein</u>, any vein that is not in the chest or abdomen

Peripheral neuroblastic tumors or pNTs: The spectrum of tumors classified by pathologists that arise from sympathetic nervous tissue; includes neuroblastoma, ganglioneuroblastoma, and ganglioneuroma.

Peripheral Stem Cells: Immature cells found circulating in the bloodstream from which new blood cells develop.

Peripherally Inserted Central Catheter (PICC Line): A long, thin, flexible tube known as a catheter. It is inserted into one of the large veins of the arm near the bend of the elbow. It is then slid into the vein until the tip sits in a large vein just above the heart.

Petechiae: Pinpoint bleeding under the skin usually caused by a low platelet count.

Pilot Study: The initial study examining a new method or treatment.

Placebo: An inert substance that resembles an actual medication, and is administered in the same way as a drug in a clinical trial.

Plasma: The clear, yellowish, fluid portion of the blood. Cells are suspended in plasma.

Platelets: One of three types of circulating blood cells that helps prevent bleeding by causing blood clots to form. Also called thrombocytes.

Platinum: A metal that is a significant ingredient of some chemotherapy drugs, such as cisplatin and carboplatin.

Ploidy: The number of sets of chromosomes in a cell or an organism. For example, haploid means one set and diploid means two sets.

Port (or "port-a-cath"): Catheter placed in chest with access just under the skin.

Positron Emission Tomography (PET) Scan: Positron emission tomography scan. A computerized representation of areas of increased glucose uptake which is used to ascertain the presence of disease.

Progressive Disease (PD): New lesion or greater than 25% increase in an existing lesion as defined by the INRC.

Prophylaxis: An effort to prevent disease.

Purging: Removing neuroblastoma cells from bone marrow or peripheral stem cells in laboratory.

R

Radiation Therapy: The use of high-energy radiation from x-ray machines, cobalt, radium or other sources for control or cure of cancer. Systemic radiation involves giving a radioactive substance, such as a radiolabeled monoclonal antibody, that circulates throughout the body.

Radioactive Iodine: A radioactive form of the chemical element iodine often used for imaging tests or as a treatment for cancer.

Radiolabeled: Any compound that has been joined with a radioactive substance.

Recurrence: The reappearance of cancer, at the same site as the original tumor or in another location, after treatment had caused it to apparently disappear.

Red Blood Cells: Cells that carry oxygen to tissues and take carbon dioxide from them. Also called erythrocytes.

Refractory: Disease that persists after intensive frontline chemotherapy.

Remission: Disappearance of the signs and symptoms of cancer. A remission is not necessarily a cure.

Resection: Surgical removal of part of an organ.

Residual Disease: Cancer cells left behind after surgery or other treatment.

Retinoid: Vitamin A or a vitamin A-like compounds such as 13-cis retinoic acid, isoretinoin or accutane.

Reverse transcription-polymerase chain reaction (RT-PCR): Method to identify a target gene and transcript level in order to detect minimal residual disease.

Risk Assignment: Neuroblastomas are divided into three risk groups based on prognostic factors determined from cytogenetics, histology, stage and age. The risk assignment determines the treatment. About 40% are low-risk, 10% are intermediate-risk, and about 50% are high-risk.

S

Sagramostin (GM-CSF): A recombinant therapeutic agent which is chemically identical to or similar to endogenous human GM-CSF. Binding to specific cell surface receptors, sargramostim modulates

the proliferation and differentiation of a variety of hematopoietic progenitor cells with some specificity towards stimulation of leukocyte production and may reverse treatment-induced neutropenias **Saline:** A solution of salt and water used for IV hydration.

Second-look Surgery: Surgery performed after primary treatment to determine whether tumor cells remain.

Sedimentation (SED) Rate: A measure of the time it takes for red blood cells to collect at the bottom of a sample of blood. It is a non-specific sign of inflammation, and can be caused by many diseases such as arthritis or wide-spread cancer. It is also called ESR.

Shimada classification: The histopathology classification of neuroblastoma tumors. The classification groups tumors based on morphological features such as degree of maturation, structure, and mitosis. Favorable histology includes tumors that are Schwannian stroma-rich, and unfavorable tumors are Schwannian stroma-poor.

Shingles: Herpes zoster, or shingles, is the reactivation of <u>varicella zoster virus</u> (<u>chickenpox</u>), producing a crop of painful <u>blisters</u>. This is common after stem cell transplants or immune suppressive therapy and is treated with acyclovir.

Shunt: A surgically created redirection of fluid from one area of the body to another.

Stable Disease: Cancer that is not decreasing or increasing in bulk or severity.

Staging: A process of determining how far a cancer has spread. Staging involves a physical exam, blood tests, x-rays, scans, and sometimes surgery. Knowing the stage of disease helps to determine treatments and prognosis.

Stem Cells: A cell from which other types of cells develop. Blood cells develop from blood-forming stem cells.

Stroma: The connective, non-functional supportive framework of a biological <u>cell</u>, <u>tissue</u>, or <u>organ</u>. Stroma-rich neuroblastomas are favorable and stroma-poor neuroblastomas are unfavorable.

Subcutaneous: Beneath the skin.

T

Teniposide: A chemotherapy drug that belongs to the family of drugs called mitotic inhibitors.

Thiotepa (ThioTEPA): A chemotherapy drug that belongs to the family of drugs called alkylating agents, used in some stem cell transplant regimens for neuroblastoma.

Thoracotomy: An operation to open the chest.

Thrombocytes: See Platelets.

Thrombocytopenia: An abnormally low number of platelets due to disease, reaction to a drug, or toxic reaction to treatments.

TNP-470: A drug that belongs to the family of drugs called angiogenesis inhibitors and blocks the growth of further blood vessels into a solid tumor.

Topoisomerase inhibitors: A substance that blocks topoisomerase enzymes, which are involved in DNA structure and cell growth, and the action of some chemotherapy drugs.

Topotecan: A <u>topoisomerase 1</u> <u>inhibitor</u> chemotherapy drug that is frequently used in relapsed neuroblastoma. In study for future use in frontline therapy.

Total Marrow Irradiation (TMI): Radiation therapy delivered to all the bones and marrow, sparing the organs. Also called TomoTherapy.

Total Parenteral Nutrition (TPN): The practice of feeding a patient intravenously, circumventing the <u>gut</u>. This is commonly required in small children undergoing intense chemotherapy and stem cell transplants.

Total-body Irradiation (TBI): Radiation therapy delivered to the whole body.

Transfusion: The infusion of components of blood or whole blood into the bloodstream.

Trimethoprim-sulfamethoxazole (Bactrim, Septra): An antibiotic used to treat infection and to prevent *pneumocystis carinii pneumonia* (pcp pneumonia).

Tumor Infiltrating Lymphocytes: White blood cells that have moved from the bloodstream and migrated into a tumor.

Tumor Marker: Substances sometimes discovered in an increased amount in the blood, other body fluids, or tissues and which may imply the presence of some types of cancer.

Tumor Necrosis Factor: A natural protein substance produced by the body, which may make tumors shrink.

Tumor Suppressor Gene: Genes in the body that can suppress or block the development of cancer.

Type and Crossmatch: To protect patients from adverse reactions to unmatched blood from blood donors, a type and crossmatch is performed to check antigens and antibodies. This process takes 45 minutes (see more).

U

Ultrasound: A study which uses high-frequency sound waves to create an image of the inside of the body.

Umbilical Cord Blood: Blood collected from the placenta at birth that contains high concentrations of stem cells needed to produce new blood cells. Parents of neuroblastoma children often are able to collect and store cord blood from later-born siblings at low or no cost.

Ureter: The tube that carries urine from the kidney to the bladder.

Urethra: The tube which empties urine from the bladder.

Urinalysis: Examination of the content of the urine.

Urokinase: A drug that dissolves blood clots or prevents them from forming.

 \mathbf{v}

Vaccine: A compound or group of compounds designed to produce an immune response to a tumor or disease.

Vancomycin: An antibiotic drug used to fight resistant bacterial infections.

Varicella: varicella-zoster virus that causes chicken pox, also one of the herpes family of viruses

Vascular: A tumor that is heavily endowed with blood vessels and thus richly supplied with blood.

Very good partial response (VGPR): Define by INRC as primary mass reduced by 90–99%, no evidence of distant disease except for skeletal residua, catecholamines normal.

Vincristine: Vinca alkaloid chemotherapy (made from the <u>Madagascar periwinkle</u>) commonly used in frontline and relapsed neuroblastoma; can cause neuropathy and loss of reflexes.

Virus: A tiny infectious agent that is smaller than bacteria. Many common infections are caused by viruses. Viruses invade cells, alter the cells' chemistry and cause them to produce more viruses. In cancer therapy, some viruses may be made into vaccines that help the body build an immune response to kill tumor cells.

W

White Blood Cells: Infection and disease fighting cells.

More general cancer terms can be found in the <u>NCI Dictionary of Cancer Terms</u> and the <u>NCI Drug Dictionary</u>. Some items used with the permission of <u>Honna Janes-Hodder</u>

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