INFORMATION ABOUT EWING SARCOMA AND PRIMITIVE NEUROECTODERMAL TUMOR

WHAT IS EWING SARCOMA?
Ewing sarcoma is a cancerous tumor primarily involving the bone (60%) of cases and soft tissues (40%). Normally, cells grow and divide to form new cells as our body needs them. When cells grow old, they die and new ones take their place. However, there are times when this orderly process goes wrong. New cells form even when the body does not need them or old cells do not die when they should.

Ewing sarcoma is the second most common bone cancer in children, adolescents and young adults. It is more common in boys than in girls. When soft tissue is affected, it is called Extraosseous Ewing sarcoma.

WHAT ARE PRIMITIVE NEUROECTODERMAL TUMORS (PNET)?
Primitive Neuroectodermal tumor or PNET is a rare cancer that also involves the bone and soft tissue. It shares many features with Ewing Sarcoma and is believed to originate from same type of cell. Ewing Sarcoma, Extraosseous Ewing sarcoma and PNET all belong to the Ewing Family of Tumors.

WHAT CAUSES EWING SARCOMA AND PNET?
The exact cause of tumors of the Ewing Sarcoma Family is unknown. There are also no known inherited or environmental risk factors identified as being associated with Ewing sarcoma and PNET. A risk factor is anything that increases a person’s chance of getting a disease. Although a chromosomal abnormality, resulting from the translocation (or exchange) of a portion between chromosome 11 and 22 has been identified in patients with these conditions, this change is not inherited and seems to occur for no apparent reason. However, this event has been shown to activate a gene that leads to the overgrowth of cells and thus the development of these cancers.

WHAT ARE THE SYMPTOMS OF EWING SARCOMA AND PNET?
The most common complaint is bone pain. This may be accompanied by swelling or a mass over the bone or soft tissue, which may be warm to touch and associated with fever. Non-specific symptoms such as weight loss and weakness may also occur. If the leg or hip bones are involved, there may pain and difficulty with walking. The physician who initially sees the child often orders X-ray films of the bone which will eventually show changes suspicious for Ewing Sarcoma. When this happens, further testing is done. When soft tissue is involved, further testing, including a biopsy or removal of tissue for microscopic examination will be performed.

HOW IS THE DIAGNOSIS OF EWING SARCOMA AND PNET MADE?
A careful health history and physical examination will be performed as well as several tests done to prove the diagnosis of Ewing sarcoma and PNET, to determine the stage and to determine if it has spread to other parts of the body.

A family history of cancer is generally asked, as well as the patient’s past illnesses and treatments.

An orthopedic surgeon (for bone involvement) or a pediatric surgeon is often consulted to perform a biopsy of the tumor. The tissue obtained will be examined by a pathologist to determine if Ewing Sarcoma or PNET is present.

Aside from a biopsy, various imaging tests may also be requested such as X-rays, a CT (Computerized Tomography) scan, an MRI (Magnetic Resonance Imaging) or a Positron Emission Tomography (PET) scan.

Other tests that may be requested include a complete blood count, baseline liver and kidney function tests, baseline hearing and heart function tests and in suspected spread of disease to other parts of the body, a CT scan of the chest, and a bone scan. A bone marrow aspiration and biopsy will also be done to determine if the bone marrow is affected by the disease.
**How does one stage Ewing Sarcoma and PNET?**

Staging refers to the process used to follow to find out if the cancer stays within the affected bone or has spread to other parts of the body. Knowing disease stage is important for treatment planning as well as knowing the child’s chance of cure.

Although there is no exact staging method employed for Ewing Sarcoma and PNET, for treatment purposes, patients are classified as having either localized (confined within the affected bone) or metastatic (with spread to other parts of the body) disease.

**How is the treatment planned and who are involved in the treatment?**

In planning the treatment for a child with Ewing Sarcoma and PNET, certain factors that impact on the chance of recovery are taken into consideration. This includes tumor location and size, whether the cancer is localized or metastatic, as well as patient age and general health.

The pediatric oncologist and orthopedic oncologist, physicians who are trained in the management of children with Ewing Sarcoma and PNET are primarily involved in the management of children when the bone is involved. In patients with soft tissue involvement, the pediatric oncologist may work primarily with a pediatric surgeon. At the same time, the expertise of other specialty physicians is required for complete care of the child (also called multidisciplinary care) - for diagnosis, treatment and supportive care. These physicians include radiologists, pediatric surgeons, oncology nurses, pharmacists, psychologists, social workers, physical and occupational therapists, etc. The members of the multidisciplinary care team all contribute to the treatment and support of the child.

**What are the available treatment options for Ewing Sarcoma and PNET?**

The treatment modalities employed for a child with Ewing Sarcoma and PNET mainly include surgery and chemotherapy and radiotherapy. Surgery is performed to remove the tumor as much as possible and to provide tissue for diagnosis (biopsy).

Chemotherapy is the use of drugs to kill cancer cells. It may be given by mouth or injected through a vein or muscle (systemic therapy). For children with Ewing Sarcoma and PNET, a combination of different drugs known to kill these specific cancer cells is given. The drugs may be given before (neoadjuvant) and/or after (adjuvant) surgery to kill remaining cells that may not be seen by any test or exam.

Radiation therapy uses high-energy X-rays or other radiation energy to kill cancer cells. The cancer cells of Ewing Sarcoma and PNET are very sensitive to radiation. Because of this, radiation therapy has become an integral part of the management of these patients. Radiation therapy is administered by a radiation oncologist.

For patients with high widespread disease or those who do not respond well to treatment, studies are now underway on the use of very high doses of chemotherapy followed by transplantation of blood forming stem cells. For these children, their own stem cells are used and is called an autologous transplant.

This information is made possible through the efforts of the Philippine Society of Pediatric Oncology, Inc. (PSPO), a subspecialty society of the Philippine Pediatric Society (PPS). For details regarding the treatment of individual patients, it is strongly recommended that they confer with their pediatric oncologist.