Neuroblastoma

Patient and family information, brought to you by the Education Committee of APSA

Overview - "What is it?"

Neuroblastoma is a type of tumor that arises from cells of the sympathetic nervous system and adrenal gland. The nervous system consists of the brain, spinal cord, and nerves. The sympathetic system is a part of the nervous system that helps control adjustments in heart, muscle, or intestinal activity, especially during stressful events.

The adrenal glands are organs located on top each kidney. The adrenal gland has cells similar to the sympathetic system that make chemicals called epinephrine (adrenaline) and norepinephrine (noradrenaline). These chemicals are important in the response to stressful situations.

Neuroblastoma is seen mostly in children. It is the third most common childhood cancer, after leukemia and brain tumors.

- More than 600 new cases are diagnosed in the United States each year.
- The average age at diagnosis is 2.5 years with 40% being diagnosed before age one.
- Neuroblastomas are associated with some congenital abnormalities and genetic diseases such as Hirschsprung disease, central hypoventilation, and neurofibromatosis.
- Since neuroblastomas arise from cells of the nervous system, it can occur in any area with sympathetic nerve tissues, including the adrenal gland (46%), the chest next to the spine (15%), the pelvis (4%), and the head and neck (3%).

Signs and Symptoms - "What symptoms will my child have?"

The symptoms of neuroblastoma vary according to the location of the main tumor and whether or not it has spread to other places in the body.

- When the tumor arises inside the belly, there may be a mass or firmness felt during a physical exam.

- When they occur in the chest, it may compress the windpipe (or trachea, which is a major airway in the neck and upper chest) and create coughing or difficulty breathing.
- If it is near the spinal cord, it can create a loss of sensation or motor function below the level of involvement.
- When spread to the lymph nodes, it can create enlarged lumps or bumps on the body, while spread to the bone marrow can create fatigue and an inability to fight infection.

Some common symptoms that children can experience with neuroblastomas regardless of its location include weight loss, fever, abdominal disturbances, diarrhea, high blood pressure, irritability, pain of bone and joints, or inability to stand up or not walk.

Diagnosis - "What tests are done to find out what my child has?"

Bloodwork: Neuroblastomas can secrete different hormones. Blood and urine samples may be tested for the presence of such hormones. When these levels are elevated, they can be used to follow the response of the disease to treatment or to watch for recurrence.

Imaging studies: Some cases of neuroblastoma can be identified during prenatal ultrasound.

- Usually when there is belly swelling, abdominal X-rays are obtained first.
- Ultrasound may be used to assess for a neck or abdominal mass.
- CT scans or MRIs may also be used to identify the mass or to check for metastasis. These types of studies give more detailed images.
- Bone scans and bone x-rays can show the presence of bone metastases.

Iodine-123-labeled metaiodobenzylguanidine (MIBG), a radiolabeled marker that is processed by the adrenal gland, is a nuclear medicine test that is useful in identifying the tumor and metastases.

If metastasis of the bone is suspected, needle aspiration of the bone marrow may be recommended.

Biopsy: A biopsy or taking a piece of tissue from the mass, may be needed to make a definite diagnosis by looking at the tissue under a microscope.

Treatment - "What will be done to make my child better?"

Staging: In all types of cancer, it is important to determine if the cancer is isolated or has it spread through the body. The treatment is dependent on the stage of the cancer.

In neuroblastoma, staging is done to look at whether the tumor can be completely removed without causing significant harm to the surrounding organs and the child. Staging will also be

done to see whether there is cancer spread in organs other than where the primary cancer is seen.

A pediatric oncologist (doctor that specializes in treating pediatric cancer) will guide you through the types of medicines and radiation to be used.

Medicine: Chemotherapy are drugs that are especially aimed at destroying neuroblastoma cells, are used either before AND after surgery or just after surgery. These are given through the vein.

- If the tumor is too big, situated in a dangerous place for the child's safety, or there is tumor spread, chemotherapy may be used to decrease the size and tumor burden before surgery to remove the tumor.
- If the tumor is too close to certain organs (close margins) or if the tumor has aggressive characteristics, chemotherapy may be needed after surgery.
- As chemotherapy attacks cells in the body that are rapidly dividing, it may also affect normal cells in the body that rapidly divide.
- Common side effects of chemotherapy include hair loss, mouth sores, loss of appetite, nausea, vomiting, diarrhea, constipation, increased chance of infections, easy bruising or bleeding and fatigue. Most of these effects are short-lived and will go away after treatment.
- Radiation therapy may also be used if any tumor cells are left behind after surgical removal of the mass.
- Radiation therapy uses high-energy rays to kill cancer cells. Although it is administered locally to the tumor, normal cells that surround the tumor may be affected as well.
- Possible short-term effects of radiation include nausea, diarrhea, fatigue, sunburn changes to the skin and hair loss.

Surgery: Due to the rare and complex nature of these tumors, treatment should be performed at centers/hospitals where surgeons are very familiar with neuroblastoma. Tumors are removed if they are able to be completely resected safely.

The mass/tumor can usually be resected at the time of diagnosis in some patients, but some have tumors that are too large to be removed right away and will get chemotherapy first.

Informed consent: A consent form is a legal document that states the tests, treatments or procedures that your child may need and the doctor or practitioner that will perform them. Before surgery, your doctor should tell you what the operation is, the goal of the surgery and other possible treatment options that are available. Your doctor should explain the risks and benefits of the surgery. You give your permission when you sign the consent form. You can have someone sign this form for you if you are not able to sign it. You have the right to understand your child's medical care in words you know. Before you sign the consent form, make sure all of your questions are answered. It is important to know that during surgery, there

are things that can happen that your doctor may have not predicted before going in. He or she will explain these to you after the surgery.

Preoperative preparation: Your child will require general anesthesia for any surgical procedure so will have to stop eating several hours before the surgery. A shower or a bath the night prior or the day of surgery helps cleanse the skin to decrease wound infections. Certain labs may be drawn to check blood count levels and to check the function of the liver.

Postoperative care: Your child will likely remain in the hospital for several days following the surgery in order to provide good pain control and intravenous fluids. Once they are eating well and able to take medications by mouth, they will be discharged.

Central line placement (Port-A-Cath® or Broviac® Catheter) will likely be necessary to give chemotherapy drugs before or after removing the tumor.

Metastases (pieces of tumor that have spread to other parts of the body, usually the lung) will need to be controlled.

Risks/Benefits:

- The main risks of surgery are bleeding and infection. Your child will likely have their blood type checked before surgery in case a blood transfusion will be necessary. They will also be given antibiotics before and maybe after surgery to help reduce the chance of infection.
- The other risk of surgery is not getting all the tumor out. This may mean that your child will need additional surgeries in the future or additional chemotherapy to rid the body of cancer cells.
- There are risks to the drugs used to treat the tumor as well and include heart and kidney problems, lowering of blood cells, developing other tumors and the risk of infection (usually from the central line).

Home Care - "What do I need to do once my child goes home?"

Diet: Your child will likely be able to resume a normal diet without restrictions.

Activity: Depending on the extent of surgery, your child might need to take it easy for a few weeks after surgery. Children tend to recover faster than adults so they may be able to return to school and light-duty activities within a week or two.

Wound care: Your surgeon should inform you of any specific wound care and whether or not you can get the incision wet. Call your surgeon if there is any redness or drainage from the incision or if your child has any fevers. You will also be given instructions in how to care for the central line.

Medicines: You may be given a prescription for pain medications. Depending on the tumor, your child may need to return to the hospital or clinic to receive chemotherapy (drugs that attack cancer cells).

What to call the doctor for: Call your surgeon for fevers (greater than 101° Fahrenheit), redness or drainage from the incision or for any vomiting or diarrhea.

Follow-up care: You will generally need to see your surgeon one to two weeks after your surgery and will also have a follow up with your oncology doctor.

Long Term Outcomes - "Are there future conditions to worry about?"

Although long-term outcomes depend on the stage of neuroblastoma and the response to treatment, many children have a good chance of long-term survival.

- Neuroblastomas that are 1) diagnosed under the age of 18 months, 2) located in the chest, neck, or lower spine and 3) do not have lymph node involvement all have more favorable outcomes. In these low-risk groups, there is a 90% long-term survival rate.
- Follow-up care after treatment for neuroblastomas is important to assess for recurrence of the tumor. Doctors will likely order follow up blood work, CT scans, PET scans, MRI or ultrasounds on a regular basis. It is important that new symptoms be reported to doctors right away.

In cases where the tumor is 1) too large, 2) there is lymph node involvement or 3) distant metastasis (spread of cancer to other organs), outcomes may not be as good and chemotherapy and/or radiotherapy may be the only option for treatment.

- Your child will require long-term follow up with the oncologist as well as the surgeon to monitor for tumor coming back and for possible side effects of the treatment.
- Long-term effects of chemotherapy depend on which drugs are used for therapy. Potential long-term side effects include hearing loss, heart damage, numbness and/or tingling, weakness, or pain. Some drugs may also increase the risk of later developing a blood cancer termed 'leukemia'.
- Long-term effects of radiation therapy include curvature of the spine, neurologic problems such as paralysis, numbness, and an inability to spontaneously void if the tumor is located on the spine. Hypothyroidism may result from radiation to the head and neck. Damage to the ovaries may occur in girls with radiation to the abdomen.

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