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Pleuropulmonary Blastoma - Childhood - Treatment Options

[1]

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ON THIS PAGE: You will learn about the different ways doctors use to treat children with this type of tumor. To see other pages, use the menu on the side of your screen.

In general, tumors in children are uncommon, so it can be hard for doctors to plan treatments unless they know what has been most effective in other children. That's why more than 60% of children with cancer are treated as part of a clinical trial. Clinical trials are research studies that compare standard treatments, which are the best proven treatments available, with newer approaches to treatments that may be more effective. Clinical trials may test such approaches as a new drug, a new combination of standard treatments, or new doses of current therapies. Studying new treatments involves careful monitoring using scientific methods, and all participants are followed closely to track their health and progress.

To take advantage of these newer treatments, all children with PPB should be treated at a specialized treatment center. Doctors at these centers have extensive experience in treating children and have access to the latest research. A doctor who specializes in treating children with cancer is called a pediatric oncologist. In many cases, a team of doctors works with a child and the family to provide care; this is called a [multidisciplinary team \[3\]](#). Pediatric cancer centers often have extra support services for children and their families, such as child life specialists, nutritionists, physical and occupational therapists, social workers, and counselors. Special activities and programs to help your child and family cope may also be available.

Descriptions of treatment options for PPB are listed below. Treatment options and recommendations depend on several factors, including the type and stage of the tumor, possible side effects, and the patient's preferences and overall health. Your child's care plan may also include treatment for symptoms and side effects, an important part of cancer care. Surgery and chemotherapy, and sometimes radiation therapy, are used to treat PPB. If all of the tumor cannot be removed during surgery, chemotherapy and/or radiation therapy may be used to kill the remaining cancerous cells.

Take time to learn about all of your child's treatment options and be sure to ask questions about things that are unclear. Also, talk about the goals of each treatment with your child's doctor and

what your child can expect while receiving the treatment. Learn more about [making treatment decisions](#) [4].

Surgery

Surgery is the removal of the tumor and surrounding tissue during an operation. It is the main treatment for PPB. A surgical oncologist is a doctor who specializes in treating a tumor using surgery. If the tumor is too large to be completely removed, or if it is not possible to completely remove the tumor, chemotherapy may be performed before surgery to reduce the size of the tumor. This is called pre-operative chemotherapy.

Side effects from surgery for PPB can vary. Talk with your child's doctor about possible short-term and long-term side effects. Learn more about [cancer surgery](#) [5].

Chemotherapy

Chemotherapy is the use of drugs to destroy tumor cells, usually by stopping the tumor cells' ability to grow and divide. Chemotherapy is given by a medical oncologist, a doctor who specializes in treating cancer with medication.

Systemic chemotherapy is delivered through the bloodstream to reach cancer cells throughout the body. Common ways to give chemotherapy include an intravenous (IV) tube placed into a vein using a needle or in a pill or capsule that is swallowed.

A chemotherapy regimen, which is your child's treatment plan, usually consists of a specific number of cycles given over a set period of time. A patient may receive one drug at a time or combinations of different drugs at the same time. Chemotherapy is often used to treat childhood tumors, and even children as young as newborn infants can be safely treated for life-threatening tumors with chemotherapy.

Chemotherapy may benefit patients with Type I PPB to reduce the risk of recurrence. Talk with your child's doctor about whether it is recommended in your child's case. Chemotherapy is always recommended for Types II and III PPB. The doctor may perform surgery first and then give chemotherapy to kill any remaining PPB cells; this is called adjuvant or post-operative chemotherapy. Sometimes, pre-operative chemotherapy is given.

Common drugs for Type 1 PPB include: cyclophosphamide (Neosar), vincristine (Vincasar) and dactinomycin (Cosmegen). Specific drugs for Types II and III often includes actinomycin D (IVADo), doxorubicin (Adriamycin), ifosfamide (Ifex), and vincristine (Vincasar).

The side effects of chemotherapy depend on the individual and the dose used, but they can include fatigue, risk of infection, nausea and vomiting, hair loss, loss of appetite, and diarrhea. These side effects usually go away once treatment is finished.

Learn more about [chemotherapy](#) [6] and [preparing for treatment](#) [7]. The medications used to treat PPB are continually being evaluated. Talking with your child's doctor is often the best way to learn about the medications prescribed for your child, their purpose, and their potential side effects or interactions with other medications. Learn more about your prescriptions by using [searchable drug databases](#)

[8].

Radiation therapy

Radiation therapy is the use of high-energy x-rays or other particles to destroy tumor cells. A doctor who specializes in giving radiation therapy to treat a tumor is called a radiation oncologist. The most common type of radiation treatment is called external-beam radiation therapy, which is radiation given from a machine outside the body. When radiation treatment is given using implants, it is called internal radiation therapy or brachytherapy. A radiation therapy regimen usually consists of a specific number of treatments given over a set period of time.

Not every child with PPB receives radiation therapy. The decision to use radiation therapy is made if the child's health care team believes that there are still cancerous cells remaining after surgery and chemotherapy.

Side effects from radiation therapy may include fatigue, mild skin reactions, upset stomach, and loose bowel movements. Most side effects go away soon after treatment is finished. However, radiation therapy can also interfere with normal bone growth and is associated with development of secondary cancers. Talk with your child's doctor about the possible short-term and long-term side effects before treatment begins. Learn more about [radiation therapy](#) [9].

Getting care for symptoms and side effects

PPB and its treatment often cause side effects. In addition to treatment to slow, stop, or eliminate the tumor, an important part of care is relieving a person's symptoms and side effects. This approach is called palliative or supportive care, and it includes supporting the patient with his or her physical, emotional, and social needs.

Palliative care can help a person at any stage of illness. People often receive treatment for the tumor and treatment to ease side effects at the same time. In fact, patients who receive both often have less severe symptoms, better quality of life, and report they are more satisfied with treatment.

Palliative treatments vary widely and often include medication, nutritional changes, relaxation techniques, and other therapies. Your child may also receive palliative treatments similar to those meant to eliminate the tumor, such as chemotherapy, surgery, and radiation therapy. Talk with your child's doctor about the goals of each treatment in the treatment plan.

Before treatment begins, talk with your child's health care team about the possible side effects of the specific treatment plan and supportive care options. And during and after treatment, be sure to tell your child's doctor or another health care team member if your child is experiencing a problem so it can be addressed as quickly as possible. Learn more about [palliative care](#) [10].

Remission and the chance of recurrence

A remission is when a tumor cannot be detected in the body and there are no symptoms. This may also be called "no evidence of disease" or NED.

A remission can be temporary or permanent. This uncertainty leads many patients and families to

feel worried or anxious that the tumor will come back. While many remissions are permanent, it is important to talk with your child's doctor about the possibility of the tumor returning. Understanding the risk of recurrence and the treatment options may help you feel more prepared if the tumor does return. Learn more about [coping with the fear of recurrence](#) [11].

If the tumor does return after the original treatment, it is called recurrent tumor. It may come back in the same place, meaning it is a local recurrence, or nearby, which is a regional recurrence. If it comes back in another place, it is a distant recurrence.

When this occurs, a cycle of testing will begin again to learn as much as possible about the recurrence. After testing is done, your child's doctor will talk with you about treatment options. Often the treatment plan will include the therapies described above, such as surgery, chemotherapy, and radiation therapy, but they may be used in a different combination or given at a different pace. Your child's doctor may also suggest clinical trials that are studying new ways to treat this type of recurrent tumor.

Additional treatment depends on where and when the tumor recurred and how it was initially treated. Surgery may be used to remove a new tumor. Chemotherapy and/or radiation therapy may also be recommended. [Bone marrow/stem cell transplant](#) [12] may also be a treatment option for some children.

When a tumor recurs, patients and their families often experience emotions such as disbelief or fear. Families are encouraged to talk with their health care team about these feelings and ask about support services to help them cope. Learn more about [dealing with recurrence](#) [13].

If treatment fails

Although treatment is successful for the majority of children with cancer, sometimes it is not. If a child's tumor cannot be cured or controlled, this is called advanced or terminal disease. This diagnosis is stressful, and it may be difficult to discuss. However, it is important to have open and honest conversations with your child's doctor and health care team to express your family's feelings, preferences, and concerns. The health care team is there to help, and many team members have special skills, experience, and knowledge to support patients and their families.

Advanced PPB is when the disease has spread beyond where it started, mostly within the chest cavity or to the diaphragm, the thin muscle under the lungs and heart that separates the chest from the abdomen. PPB can also spread through the bloodstream to other organs, especially the bones, liver, and brain.

Areas where PPB has spread are found by imaging tests, such as a CT scan, MRI scan, or bone scan. See the [Diagnosis](#) [14] section for more information on these tests. PPB that has spread is rarely treated with surgery; other treatments will be recommended. If one area where PPB has spread is found on a scan, it is likely that the PPB has spread to other areas that cannot be seen on the scan.

Parents or guardians are encouraged to think about where the child would be most comfortable: at home, in a home-like setting elsewhere, in the hospital, or in a hospice environment. Hospice care is a type of palliative care for people who are expected to live less than six months. It is

designed to provide the best possible quality of life for people who are near the end of life. Nursing care and special equipment can make staying at home a workable alternative for many families. Some children may be happier if they can arrange to attend school part-time or keep up other activities and social connections. The child's health care team can help parents or guardians decide on an appropriate level of activity. Making sure a child is physically comfortable and free from pain is extremely important as part of end-of-life care. Learn more about [caring for a terminally ill child](#) [15] and [advanced cancer care planning](#) [16].

The death of a child is an enormous tragedy, and families may need support to help them cope with the loss. Pediatric cancer centers often have professional staff and support groups to help with the process of grieving. Learn more on [grieving the loss of a child](#) [17].

The next section helps explain clinical trials, which are research studies. Use the menu on the side of your screen to select About Clinical Trials, or you can select another section, to continue reading this guide.

Links:

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- [2] <http://www.cancer.net/about-us>
- [3] <http://www.cancer.net/node/25356>
- [4] <http://www.cancer.net/node/24582>
- [5] <http://www.cancer.net/node/24720>
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