Hepatoblastoma (Liver Tumor)

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

Overview - "What is it?"

Hepatoblastoma is rare but is the most common liver cancer tumor in children.

There are approximately 100 new cases of hepatoblastoma per year, or 1.6 children per million per year. The incidence of hepatoblastoma continues to rise yearly, but the reason remains unknown. Children typically present before age 4, with best prognosis when the child is less than 2 years old.

Multiple risk factors have been suggested for developing hepatoblastoma:

- Extreme prematurity and low birth weight (<1500 g)
- Tumor syndromes: Familial adenomatous polyposis and Gardner syndrome, Beckwith-Wiedemann, and Li-Fraumeni
- Sotos syndrome
- Simpson-Golabi-Behmel syndrome
- Glycogen storage disease

Genetic diseases: Trisomy 18 and 21

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

Most commonly children present with a painless lump or mass felt on the right side or middle portion of the belly, found either by parent or physician.

Most children are asymptomatic or show no symptoms of the tumor. However, if children do have symptoms, they may include poor appetite, jaundice (yellowing of the skin), or itching.

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

Physical examination by a doctor. Typically, a lump or mass can be felt on exam in the right side of the abdomen.

Blood tests: if a liver tumor is suspected, routine labs to check for blood cell counts, liver enzyme levels and tumor markers, specifically alpha-fetoprotein (AFP) will be obtained

Abdominal Ultrasound: uses sound waves to create images and pictures. It is typically the first test to evaluate abdominal masses that are felt on exam, because it does not use radiation, it is cost effective, and does not require sedation for young children.

Computed tomography (CT) scans: uses radiation. Provides a detailed look at the liver mass and its relationship to other organs, blood vessels, and gives important information. A CT may be performed of the chest to look for tumor in the lungs or outside of the liver.

MRI (Magnetic Resonance Imaging): Does not have radiation, but is typically a longer imaging examination. Most younger children will require anesthesia for this test. Similar to CT scans, MRIs give better detail of the liver mass location and relationship with important surrounding structures.

Liver tumor biopsy or taking a piece of tissue from the mass is common to make a definite diagnosis by looking at the tissue under a microscope and help with planning therapy. There are different types of hepatoblastoma which are confirmed by biopsy.

Other conditions can mimic hepatoblastoma: Non-cancerous liver tumors/masses such as benign hemangiomas (blood vessel abnormality) or hamartomas can look like this tumor. Other cancerous masses arising from the liver are hepatocellular carcinoma (HCC), germ cell tumors (infantile choriocarcinoma of the liver), undifferentiated embryonal sarcoma of the liver. Labs tests, imaging, and biopsy all together help tell the difference.

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

Treatment is multifactorial. Due to the rarity of hepatoblastoma, your child's treatment should take place at an institution/hospital with doctors who have more experience with the disease. Each child's treatment plan is formulated with the input of many doctors: pediatric oncologist (doctor specializing in treatment of cancer), pediatric surgeon, pathologist, radiologist, to name a few.

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

- In hepatoblastoma, staging is done to look at whether the tumor can be completely removed from the liver and leave enough liver to have function. Staging will also be done to see whether there is cancer spread in organs other than the liver.

- A pediatric oncologist will guide you through the types of medicines and other interventions to be used.

Medicine: Chemotherapy are medications given through an IV in a large vein aimed at destroying hepatoblastoma cells. These medications can be used before AND after surgery, or only after surgery. If the tumor is too large to be removed with surgery at diagnosis, chemotherapy will be given prior to surgery.

Surgery: Removing all the tumor is the goal of all therapies. Tumors are surgically removed if the entire tumor can be removed while leaving the patient with enough liver to function and support the patient. Occasionally, if the tumor is too large or involves important structures, patients will require a liver transplant.

Preoperative preparation: Your child will require general anesthesia for any surgical procedure, so he/she 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 remain in the hospital for several days following the surgery to provide good pain control and intravenous fluids. He or she may stay in the Intensive Care Unit (ICU) after surgery for a day or two. 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) may be necessary to give chemotherapy drugs before or after removing the liver tumor.
- Metastases (pieces of tumor that have spread to other parts of the body, usually the lung) may require removal by surgery as well.

Risks:

- The main risks of surgery are bleeding and infection. Your child's blood type will be checked before surgery because blood transfusions are at times necessary. They will be given antibiotics before and possibly after surgery to help reduce the chance of infection.
- Other risks of surgery may include: bile leak or not removing all the tumor. This may mean your child will need additional surgeries in the future or additional chemotherapy to rid the liver of cancer cells.
- Chemotherapy drugs have risks as well and including 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 should 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 how to care for your child's incision and whether it can get wet. Call your surgeon if there is redness or drainage from the incision or if your child experiences fevers. You will be given instructions on how to care for the central line, if you are sent home with one.

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.

Patients are on chemotherapy may be asked to avoid acetaminophen (Tylenol®) or ibuprofen (Motrin® or Advil®). Check with your doctors on recommendations on pain medications.

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 typically 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?"

The overall survival of children with hepatoblastoma has been steadily improving over the last few decades. The prognosis or chance your child will do well is dependent on success of removing the tumor during surgery and how well he/she responds to the chemotherapy.

Your child will require long-term follow up with the oncologist and the surgeon to monitor for tumor coming back, and for possible side effects of treating the tumor. Your child's blood will be checked periodically and may require imaging studies such as CT scans or MRIs.

If the tumor cannot be completely removed or returns after treatment, a liver transplant may be necessary. At that point, follow-up with a transplant surgery team may be arranged.

Updated: 11/2021
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