The Stephan L. Harris Center for Chordoma Care

The Stephan L. Harris Center for Chordoma Care, located at Massachusetts General Hospital in Boston, MA, is among the most advanced & comprehensive chordoma treatment centers in the world. Established by a generous gift from Mrs. Stephan L. Harris, in memory of her husband, the Stephan L. Harris Center for Chordoma Care is dedicated to providing the most innovative compassionate care for patients and families facing the challenging diagnosis of chordoma.

617-724-3700
www.massgeneral.org/harris-center

THE STEPHAN L. HARRIS CENTER FOR CHORDOMA CARE

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Thomas F. DeLaney, MD
Medical Director

Joseph H. Schwab, MD, MS
Surgical Director

Anne M. Fiore, DNP, ANP-BC
Nursing Director

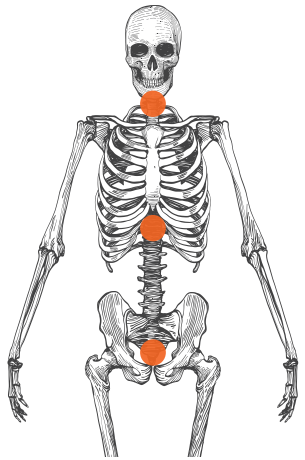
Our dedicated chordoma team includes:

- Radiologists
- Pathologists
- Medical Oncologists
- Radiation Oncologists
- Surgeons & Advanced Practitioners:
  - Orthopaedic Oncology
  - Spine Oncology
  - Surgical Oncology
  - Neurosurgery
  - Plastic & Reconstructive Surgery
  - Thoracic Surgery
  - Urology Surgery
  - Vascular Surgery
- Physical Medicine & Rehabilitation Physicians

Our team also includes: physical therapists, occupational therapists, social workers, nutritionists and ostomy nurses.

CHORDOMA: A RARE TUMOR

Chordomas are rare, locally aggressive bone tumors (neoplasm). They account for about 3% of all bone tumors and about 20% of primary spinal tumors. In adults, 50% of chordomas involve the sacral region, 35% occur at the base of the skull and 15% occur at other sites in the spine.



35% OF CASES IN THE BASE OF THE SKULL

15% OF CASES IN THE VERTEBRAL BODIES

50% OF CASES IN THE BASE OF THE SPINE

The National Cancer Institute classifies a tumor as rare when there are less than 45,000 cases per year. About 300 patients are diagnosed with chordoma each year in the United States, and about 700 in all of Europe. At any given time, fewer than one in 100,000 people are living with chordoma.

Chordomas are complicated tumors to treat because of the involvement of adjacent critical structures such as the brain stem (with clival chordomas), vertebral bodies and spinal cord (with cervical, thoracic and lumbar chordomas), and important nerves that control bowel, bladder and sexual function (with sacral chordomas). The tumors are generally slow-growing and can affect adjacent organs, tissues and bones. Chordomas may develop at any age but usually occur between 40 and 70 years of age.

Most patients with chordomas present with pain in the location of the tumor, along with changes in sensation or function. Chordomas located at the base of the skull may cause headaches, neck pain or double vision. Thoracic and lumbar chordomas may cause numbness or weakness in the arms or legs. Sacral chordomas can cause bowel, bladder or sexual dysfunction.

There are no known environmental, dietary, or lifestyle risk factors that cause chordoma. The majority of chordomas occur at random and are not a direct result of an inherited genetic trait. Treatment for chordoma must be personalized, with each person's condition carefully considered.

UNDERSTANDING CHORDOMA

Biopsy and Pathology

Biopsies are done to determine the chordoma diagnosis. Ideally, the biopsy is done at a center of excellence experienced in the management of chordoma.

Chordoma tumors develop from cells called the notochord, which are similar to cartilage tissue in an embryo that helps develop the spine. The notochord disappears when the fetus is about eight weeks old, but some notochord cells remain behind in the bones of the spine and skull base forming tumors, either benign notochordal cell tumors (BNCT) or very rarely, malignant chordoma.

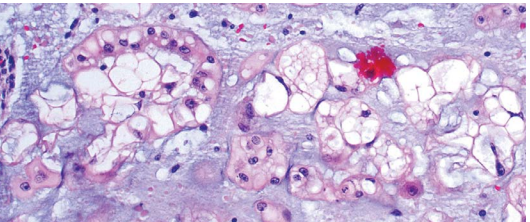
There are four subtypes of chordoma:

- **Conventional (or classic) chordoma** is the most common form.
- **Poorly differentiated chordoma** can be more aggressive and faster growing than conventional chordoma. It is more common in pediatric and young adult patients, and those with skull base (clival) and cervical patients.
- **Dedifferentiated chordoma** is the most aggressive and fastest growing chordoma. It is more likely to metastasize than conventional chordoma.
- **Chondroid chordoma** is a term used in the past when it was difficult for pathologists to distinguish conventional chordoma from chondrosarcoma (malignant bone tumor of cartilage tissue). This is no longer a problem because nearly all conventional chordomas express brachyury, unlike cartilage tumors like chondrosarcoma that do not express brachyury. Brachyury is a protein, which regulates other genes that are important in embryonic development. Brachyury is highly expressed in nearly every chordoma.

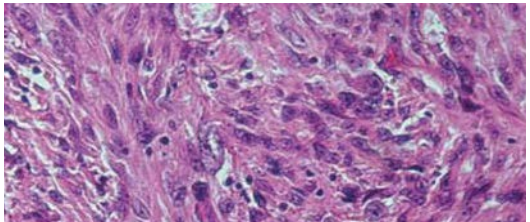
"Brachyury is expressed in all notochordal cell tumors, including chordomas. Brachyury is a diagnostic marker, but it does not distinguish between benign notochordal cell tumors (BNCT) and malignant chordoma. It is important to distinguish between BNCT and chordoma because BNCT does not require treatment, just monitoring. It is imperative that these cases be evaluated by an experienced bone pathologist and musculoskeletal radiologist."

– G. Petur Nielsen, MD

Conventional chordoma



Dedifferentiated chordoma



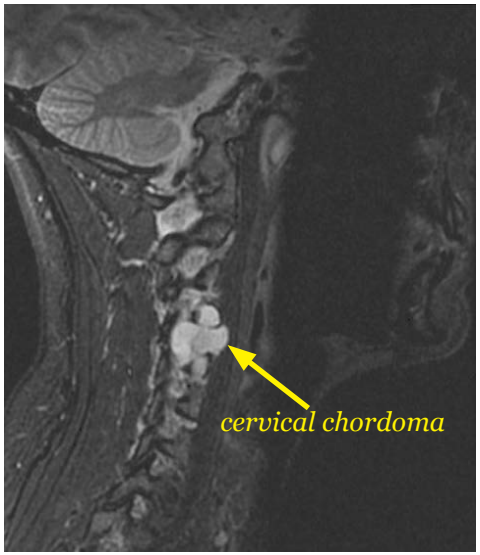
OUR CULTURE OF CARE

Personalized care is the cornerstone of treatment at the Stephan Harris Chordoma Center. Each patient is evaluated and cared for by a multidisciplinary team of experts who specialize in the diagnosis, treatment and care of patients with chordoma. As part of Mass General, the largest teaching hospital of Harvard Medical School, patients have access to vast resources and services that only a preeminent academic medical center can offer.

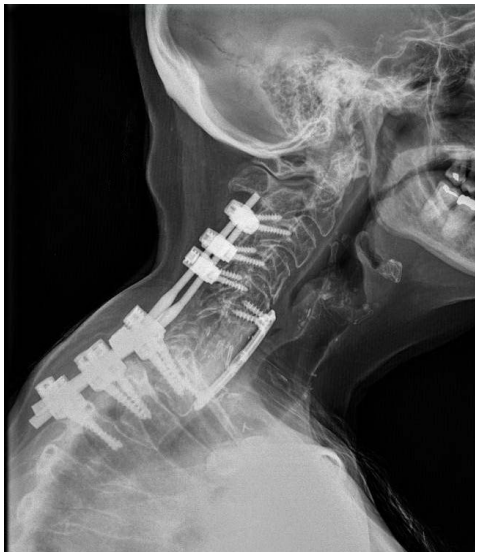
Pioneering care is provided by a close-knit clinical team that includes physicians in radiation, medical, surgical and orthopaedic oncology, neurosurgery, plastic reconstructive surgery, radiology and pathology as well as nurse practitioners, nurses, cancer rehab physicians, physical therapists, occupational therapists, nutritionists and social workers who specialize in cancer care. Other surgical specialties such as thoracic, urology and colorectal surgery are involved per individual needs.

Patients and families are seen in our multidisciplinary center where all aspects of care are managed in one location. To ensure high-quality seamless care, the entire medical team meets weekly to discuss each patient's case, reviewing current imaging studies and pathology and recommending the most effective treatment plan.

It is important to remember that the prognosis for each chordoma patient is different, and depends on many factors such as the patient's age, subtype of chordoma, size and location of the tumor, previous surgeries and treatment, extent of the surgical resection and involvement of adjacent organs, tissues, nerves and bone.



MRI showing cervical chordoma



Cervical reconstruction

BEST SCIENCE, BEST CARE

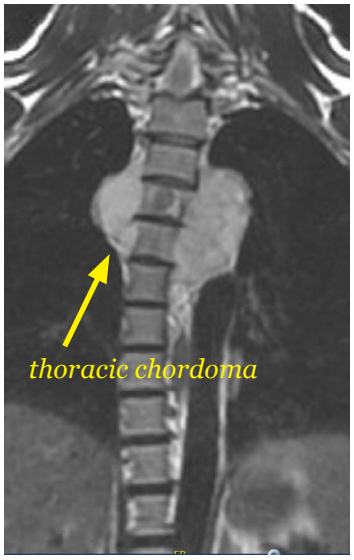
Exceptional and compassionate care are hallmarks of the Harris Chordoma Center. This care begins with groundbreaking science. Our clinicians are at the forefront of both basic science and clinical research to discover better ways to diagnose, treat and manage the disease. This research has resulted in a unique treatment plan at Mass General for patients with chordoma.

While traditional treatment involves surgery alone, our team focuses on the combination of surgery and radiation to help prevent the tumor from recurring at the original site. This approach has proven to be extremely effective in ensuring the best outcomes and preventing the spread of disease. In fact, a recent published study showed a four-year survival rate among 87% of the patients who received this treatment, provided surgery was performed for the first time at Mass General.

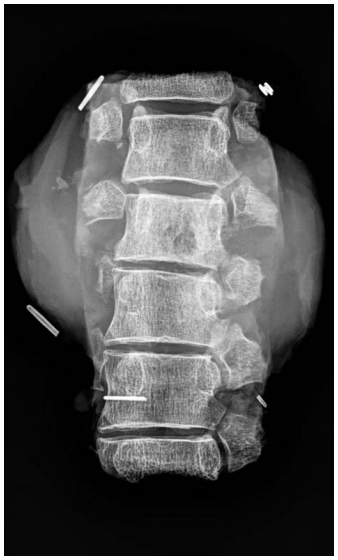
The Stephan Harris Chordoma Center's growing patient database offers extensive opportunities for analysis and discovery. In addition, ongoing clinical trials test new therapies that enable our researchers to better understand and treat chordoma, which offers additional hope for cure.

"We are world leaders in clinical trials for chordoma patients. We are exploring targeted and immunotherapies to help find new treatments for this disease."

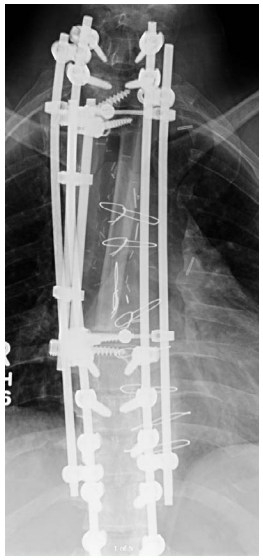
- Gregory M. Cote, MD, PhD & Edwin Choy, MD, PhD



MRI showing thoracic chordoma



Chordoma tumor & spine removed during resection



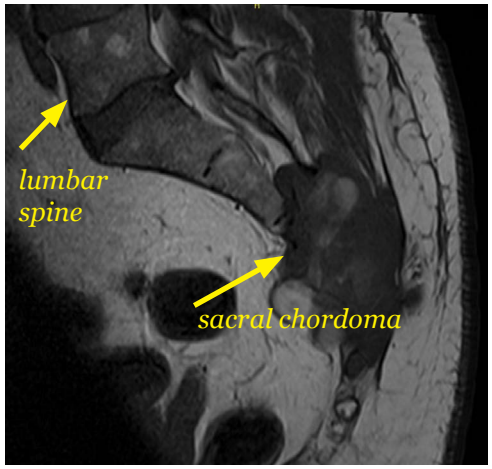
Thoracic reconstruction

One of the new advances in chordoma treatment is immunotherapy. The development of checkpoint inhibitors takes advantage of the patient's (host) immune response. The essential role of the immune system is to detect normal cells versus those that appear foreign. This lets the immune system attack the foreign cells while leaving the normal cells alone. To do this, it uses "checkpoints," which are molecules on certain immune cells that need to be activated (or inactivated) to start an immune response. Cancer cells are smart - they can find ways to use these checkpoints to avoid being attacked by the immune system, but new treatments with immunotherapy target these checkpoints. Immunotherapy has transformed the treatment landscape of oncology, although its role for treatment of chordoma is still being investigated.

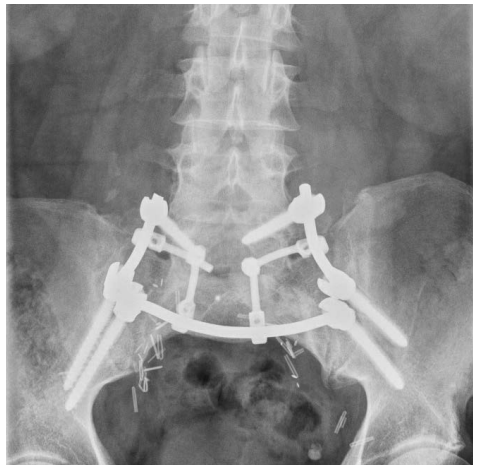
In addition, the chordoma vaccine trial currently active at Mass General Hospital holds promise for our patients. Our laboratory is focusing on how the host's immune system screens its cells for signs of cancer using immune surveillance. One of the ways in which tumors escape immune surveillance is by suppressing their expression of important proteins on the surface of the cancer cells. These proteins are known as HLA class 1 antigens.

"Our laboratory is investigating the interplay of the HLA class I antigens in chordoma and the host's immune system. The insight we gain from these interactions will help us design clinical trials to improve chordoma immunotherapy treatment."

– Joseph H. Schwab, MD, MS, Surgical Director



MRI showing sacral chordoma



Total sacrectomy

WHAT CAN I EXPECT?

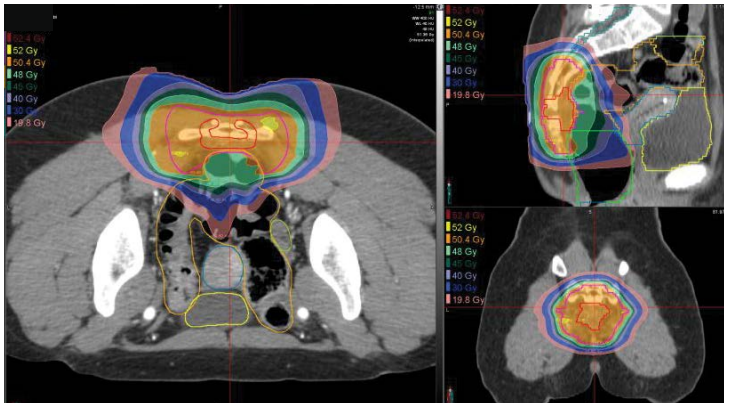
Initial Visit:
You will meet the multidisciplinary chordoma team (surgeon, medical oncologist, radiation oncologist, nurse practitioners, fellows, residents). Determining a diagnosis and personalized treatment plan can cause anxiety - your chordoma team will support you through this process.

Biopsy:
If you had a biopsy at another hospital, our pathologists will review your slides to confirm the diagnosis. If you have not had a biopsy, you will undergo a CT-guided needle biopsy with one of our musculoskeletal radiologists.

Imaging:
All outside imaging is reviewed by our musculoskeletal radiologists. Additional scans are ordered to complete the initial chordoma work-up (MRI of the entire spine; CT of chest, abdomen and pelvis; MRI of exact tumor location).

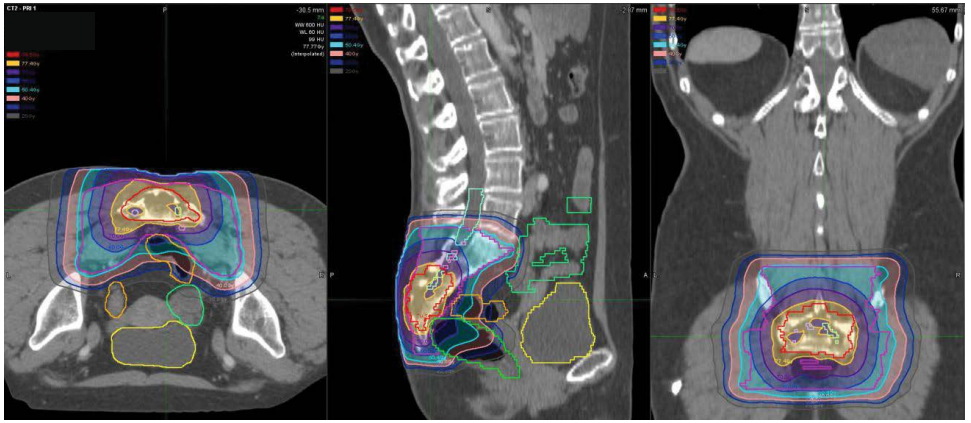
Personalized Treatment Plan:
Once the diagnosis is confirmed and all scans are reviewed, you will have a lengthy visit with your chordoma team to discuss all treatment options. With your team, you will decide which treatment option is suitable for you, your quality of life and your family.

1. **Preoperative radiation + surgery + postoperative radiation**
   - Daily radiation (Monday-Friday) for five weeks.
   - Treatment break for one month to allow radiated skin to recover.
   - 1-3 scheduled procedures are necessary to remove the tumor in total and reconstruct the defect left behind - called "staged surgeries."
     - Reconstruction of the defect can involve use of a metal cage, allograft strut, vascularized fibula, spinal instrumentation and various types of muscle flaps.
   - Postoperative daily radiation once surgical incisions have healed.



Preoperative radiation for sacral chordoma, total dose 50.4 Gy, followed by surgical resection, with plan for postoperative boost to tumor bed, dose to be based on surgical margins.

2. **Definitive Radiation (which means radiation only)**
   - Daily radiation (Monday-Friday) for seven - eight weeks.



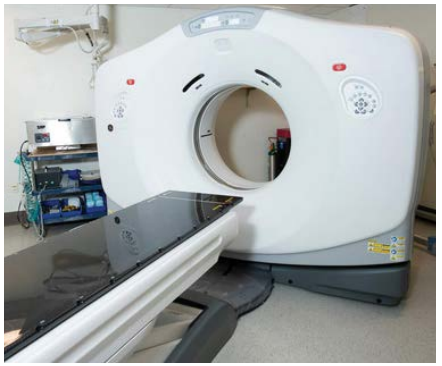
Definitive radiation plan for sacral chordoma (radiation only, no surgery). Total dose 77.4 Gy

"Successful treatment of chordoma requires high radiation doses while sparing adjacent uninvolved normal tissue. We use sophisticated radiation techniques including protons and dural plaque brachytherapy to maximize cure rate and minimize the risk of treatment-associated side effects."

– Thomas F. DeLaney, MD, Medical Director

Most patients regain their strength and endurance over 12 months. The chordoma care and survivorship continuum is life-long with oncologic surveillance visits as follows: every three months for two years; every six months for three years; followed by annual visits.

Cure is possible, but in some cases, chordomas can recur (local recurrence) after treatment, usually in the same place as the initial tumor. In some cases, chordoma can spread to the lungs, liver, other bones or soft tissue (metastatic disease). Surveillance after treatment is required for at least 10 years.



LIVING WITH THE AFTERMATH OF CHORDOMA TREATMENT

All aspects contributing to a patient's quality of life – physical, psychological, social and spiritual – can be impacted by chordoma treatment.

A malignant tumor diagnosis is anxiety provoking. All patients react and cope differently to their diagnosis, treatments and side-effects of surgery due to the chordoma size and location in the body. The goal of the chordoma team is to eradicate feelings of loss and isolation during transition to home life, the most vulnerable time of their surgical journey (per patient report). We will encourage self-management by providing practical support, resources and strategies to manage daily activities.

Maintaining daily activities of living during and after chordoma treatment may be difficult, including hygiene (bowel/bladder function), work responsibilities, school studies, as well as social and sexual relations.

Complex chordoma surgeries and/or definitive radiation can cause life-altering changes. Patients and their families require adequate time to adjust, adapt and cope with a new lifestyle:

- **Physical:** fatigue, pain, neuropathy, lymphedema, limited mobility, bowel/bladder/sexual dysfunction, inability to independently fulfill daily activities

"No matter who you are, you're going to feel depressed because it's just a total change in your body and how you feel. I had to sit in a chair to shower, have somebody help me dress, couldn't put my socks or do basic stuff that regular people do. I thought I'd just be back to normal. Permanent changes make you lose how you feel."

– patient with lumbar chordoma

- **Psychological:** emotional distress, withdraw from family/friends, anxiety, depression, cognitive changes, post-traumatic stress symptoms, fear of recurrence, ineffective coping strategies

"I learn something new about myself everyday - about my resilience and spirituality. I take nothing for grated anymore . . . it's a whole different experience, a positive one. This whole thing has been a blessing and a curse."

– patient with sacral chordoma

- **Social:** Financial worries, relationship challenges, altered familial roles, work issues/disability

"My wife is amazing; she's my rock; my solar panel; she recharges me; I get energy from her and I like to think she gets energy from me. This cancer made us even closer."

– patient with thoracic chordoma

- **Spiritual:** loss of faith, questioning purpose in life, asking "why me"

"I'm almost glad I had this cancer because just living my day-to-day life I have this unshakable calm happiness - happy to do groceries, lift my bag, drink coffee again, go to work - just the most mundane things are so pleasant and an appreciation that you cannot gain any other way. It's a gift. It does feel like a gift, and you're part of a special club of people that have done this, seen this, and know what it is."

– patient with cervical chordoma

Strategies to help you live with the aftermath of chordoma treatment include:

- various medications (narcotic and non-narcotic), nerve blocks and other innovative procedures for pain control
- supportive oncology such as: social services, palliative care, nutrition, ostomy care
- psychobehavioral strategies such as: psychiatry and psychotherapy, mindfulness techniques, meditation, guided imagery, and other integrative therapies (massage, acupuncture, music and animal therapy, yoga, reiki)

Your team will be with you every step of the way along your chordoma care journey. Whatever your specific needs are, the team will:

LISTEN to you and your family

UNDERSTAND your unique needs and

ACT to help you live with the aftermath of treatment.

"Patients with chordoma need a team of highly skilled, experienced clinicians to meet the challenges of this complex disease. Few centers exist where these rare tumors are treated. The initial intervention in managing chordoma is our patients' best chance at obtaining a positive outcome."

-Anne M. Fiore, DNP, ANP-BC, Nursing Director

PATIENT REPORTED OUTCOMES

When patients tell us how they are doing after surgery and/or radiation relative to their physical and mental health, this is called **patient reported outcomes.**

Dr. Schwab and his research team were the first group to conduct, report and publish patient reported functional outcomes for malignant tumors of the spine and pelvis. During your chordoma journey, we will ask you to complete research questionnaires. Your answers regarding your physical and mental health function are translated into percentages, which are compared to normal data from quality-of-life outcome surveys of the National Institute of Health's Patient Reported Outcome Measurement Information System (PROMIS). Such data are valuable for educating patients on expectations of postoperative function and help us to prepare future patients undergoing the same treatment. Your participation in completing these research surveys is greatly appreciated because you are helping future chordoma patients.

DNP Fiore's qualitative research uncovered patients' true thoughts and feelings about their radiation and surgery treatment. This research complimented Dr. Schwab's quantitative research, which revealed surgical risk percentages. For example, a percentage (quantitative research) representing the risk of bowel, bladder and sexual dysfunction after sacral resection was graphically described by patients in their everyday life (qualitative research). These patient responses provided a uniquely humanizing perspective into the meaning of these risk percentages.

"I know the physical limitations as a result of surgery, and I know how I want to push those limitations, but I also want to productive and have no pain . . . I have a very different body, and I am trying to understand that body."

– patient with thoracic chordoma

Patient reported outcomes, family-centric care and shared decision making became healthcare's major priorities after the passing of the Affordable Care Act (Obamacare) in 2010.

- **Patient-family centered care** comes naturally to the dedicated chordoma team. Your family and friends are included from the initial visit through your care continuum. Research shows the positive impact that support from family and friends has on cancer patients.
- **Shared decision making** is fostered at the time of your initial consultation with the chordoma team. You have the final say regarding your treatment plan because you are the one challenged to adapt your lifestyle and cope with the aftermath of life-altering surgeries and radiation treatment for chordoma.

RESOURCES

Chordoma-specific resources:

- *Stephan Harris Chordoma Center:* www.massgeneral.org/harris-center
- *Chordoma Foundation:* www.chordomafoundation.org

Resources for all types of cancer:

- *National Comprehensive Cancer Network (NCCN):* www.nccn.org
- *American Society of Clinical Oncology:* www.cancer.net
- *American Cancer Society:* www.cancer.org
- *National Cancer Institute:* www.cancer.gov
- *National Library of Medicine at the National Institutes of Health clinical trial database:* www.clinicaltrials.gov

Mass General Cancer Center resources:

Cancer can take a dramatic physical toll on patients' bodies, but the mental, emotional and financial demands of serious illness can be just as difficult. The support programs at Mass General can help patients and their families cope with the challenges of a cancer diagnosis and treatment.

- *Supportive Care Services*
  - www.massgeneral.org/cancer/experience/supportive.aspx
- *Workshops & Support Groups*
  - www.massgeneral.org/cancer/supportservices/groups.aspx
- *Integrative Therapies*
  - www.massgeneral.org/cancer/supportservices/integrative.aspx
- *Housing Accommodations for Mass General Patients & Families*
  - www.massgeneral.org/visit/boston

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Where best science and best care create hope.

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