National Cancer Institute

A Snapshot of

Sarcoma

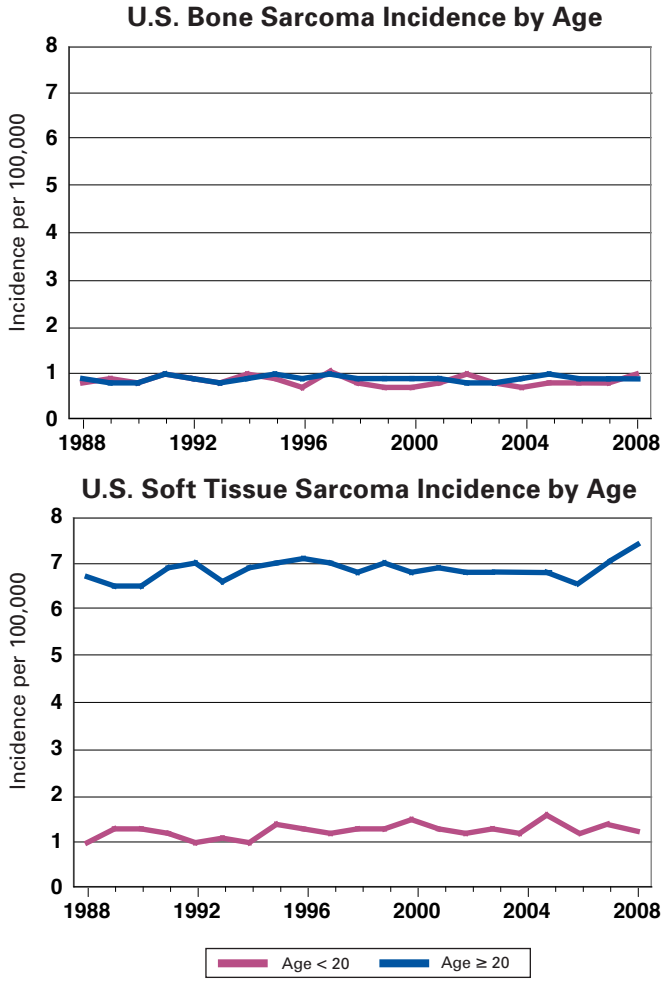
Incidence Rate Trends

Sarcomas are a diverse and relatively rare group of malignant tumors that develop in soft tissue and bone. Soft tissue sarcomas form in fat, muscles, nerves, joints, blood vessels, and deep skin tissues. Osteosarcomas and Ewing sarcomas form in bone and cartilage. Sarcomas can be difficult to distinguish from other malignancies when they are found within organs; thus, their incidence is probably underestimated. Because sarcomas are more common in adolescents and young adults than most other cancers, the number of years of life lost to sarcomas is substantial despite their relatively low incidence. It is estimated that in 2011, approximately 10,980 and 2,810 Americans will be diagnosed with soft tissue and bone sarcomas, respectively, and that approximately 3,920 and 1,490 people, respectively, will die from these diseases.

Soft tissue sarcoma[1] and osteosarcoma (bone sarcoma) incidence rates have remained relatively constant over the past 30 years; however, soft tissue sarcoma is more deadly, possibly because the lack of specific symptoms at early disease stages may lead to delays in diagnosis. Several subtypes of both osteosarcoma and soft tissue sarcoma exist; the exact number of Americans with each sarcoma subtype is unknown.

Source for incidence data: Surveillance, Epidemiology, and End Results (SEER) Program and the National Center for Health Statistics. Additional statistics and charts are available at http://seer.cancer.gov/.

[1] Does not include Kaposi sarcoma, which is addressed in a separate Snapshot.



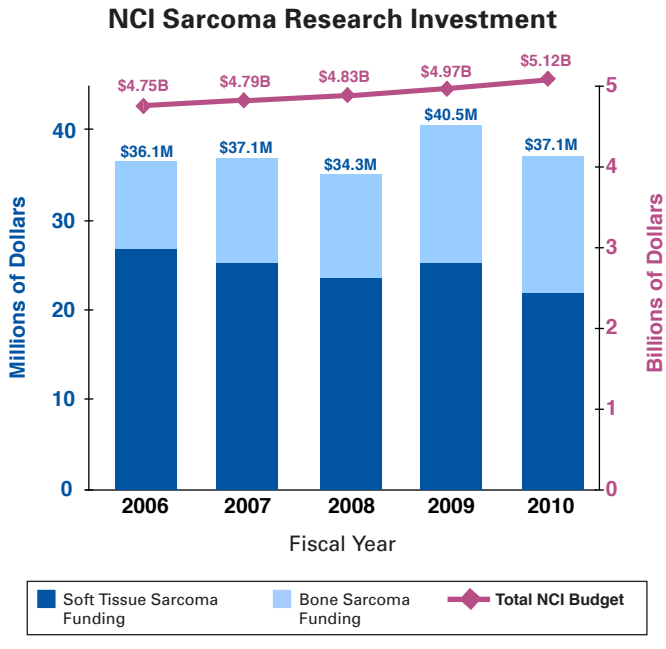
Trends in NCI Funding for Sarcoma Research

Funding data for sarcoma research have been collected only since 2005. The National Cancer Institute's (NCI) investment[2] in sarcoma research increased from $36.1 million in fiscal year (FY) 2006 to $37.1 million in FY 2010. In addition, NCI supported $22.6 million in sarcoma research in FY 2009 and 2010 using funding from the American Recovery and Reinvestment Act (ARRA).[3]

Source: NCI Office of Budget and Finance (http://obf.cancer.gov).

[2] The estimated NCI investment is based on funding associated with a broad range of peer-reviewed scientific activities. For additional information on research planning and budgeting at the National Institutes of Health (NIH), see http://www.nih.gov/about/.

[3] For more information regarding ARRA funding at NCI, see http://www.cancer.gov/aboutnci/recovery/recoveryfunding.



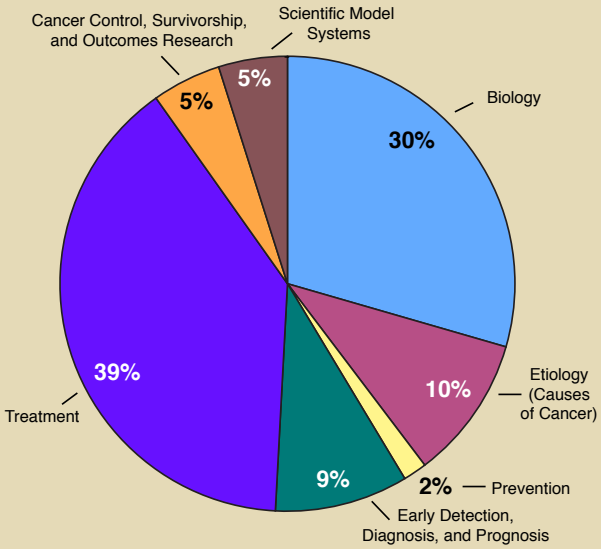
U.S. DEPARTMENT OF HEALTH AND HUMAN SERVICES

National Institutes of Health

Examples of NCI Activities Relevant to Sarcoma

- The **Prevention Agents Program** provides scientific and administrative oversight for chemoprevention agent development from preclinical research to phase I clinical trials. The program is currently supporting research on several agents with the potential to block, reverse, or delay sarcoma. http://prevention.cancer.gov/programs-resources/groups/cad/programs/agents
- NCI's **Drug Development Group** supports preclinical/clinical development of therapeutics, including potential agents for the treatment of sarcoma. http://dtp.nci.nih.gov/docs/ddg/ddg_current.html
- NCI's **Strategic Partnering to Evaluate Cancer Signatures (SPECS)** program supports large collaborative research groups that explore how information from molecular studies can be used to improve the care and outcomes of cancer patients. One SPECS project is refining and validating molecular signatures to provide a more accurate diagnosis of childhood sarcomas and better predict their clinical behavior. http://dctd.cancer.gov/ProgramPages/cdp/major_initiatives_strategic_partnering.htm
- Phase I **Study of Trabectedin in Pediatric Patients with Relapsed or Refractory Solid Tumors** is an early-phase clinical trial of a new drug called trabectedin in children; the drug is also being developed for the treatment of adult patients with soft tissue sarcoma, ovarian cancer, and other solid tumors. http://www.cancer.gov/aboutnci/ncicancerbulletin/archive/2009/063009/page8
- The **Childhood Cancer Therapeutically Applicable Research to Generate Effective Treatments (TARGET)** initiative is identifying and validating therapeutic targets to develop new, more effective treatments for children with pediatric cancers, including osteosarcoma. http://target.cancer.gov/
- The first sarcoma-specific **Specialized Program of Research Excellence (SPORE)** is developing therapies targeted to the molecular, genetic, epigenetic, and signaling pathway alterations that are specific to sarcoma type and subtype. http://trp.cancer.gov/spores/sarcoma.htm

NCI Sarcoma Research Portfolio



Percentage of Total Dollars by Scientific Area
Fiscal Year 2010

Data source: The NCI Funded Research Portfolio. Only projects with assigned scientific area codes are included. A description of relevant research projects can be found on the NCI Funded Research Portfolio Web site at http://fundedresearch.cancer.gov

- The **Soft Tissue Sarcoma Fact Sheet** contains information about the possible causes of soft tissue sarcoma; its frequency, symptoms, diagnosis, and treatments; as well as ongoing clinical trials. Information specialists can also answer questions about cancer at 1-800-4-CANCER. http://www.cancer.gov/cancertopics/factsheet/Sites-Types/soft-tissue-sarcoma
- The **NCI Soft Tissue Sarcoma Home Page**, **Bone Cancer Home Page**, **Uterine Sarcoma Home Page**, and **Ewing Family of Tumors Home Page** provide up-to-date information on treatment, prevention, genetics, screening, and testing for various types of sarcoma. http://www.cancer.gov/cancerinfo/types/soft-tissue-sarcoma/, http://www.cancer.gov/cancertopics/types/bone, http://www.cancer.gov/cancerinfo/types/uterinesarcoma/, and http://www.cancer.gov/cancertopics/types/ewing

Selected Advances in Sarcoma Research

- A genome-wide association study has **identified a set of genes potentially regulated by a mutated protein associated with** an aggressive form of rhabdomyosarcoma, a type of soft tissue sarcoma; these genes may represent new therapeutic targets for this disease. http://home.ccr.cancer.gov/inthejournals/cao.asp and http://www.ncbi.nlm.nih.gov/pubmed/20663909
- Researchers have demonstrated the **first successful use of adoptive immunotherapy** to treat patients with synovial cell sarcoma in a small phase II clinical trial. http://www.cancer.gov/ncicancerbulletin/020811/page3#c and http://www.ncbi.nlm.nih.gov/pubmed/21282551
- Results from a phase II clinical trial show that an **experimental drug called cediranib may benefit patients with alveolar soft part sarcoma**, a rare slow-growing cancer. http://www.cancer.gov/newscenter/pressreleases/2011/SoftTissueSarcomaASCO2011
- Researchers have isolated a subpopulation of Ewing sarcoma cells that have characteristics consistent with cancer stem cells; targeting these cells may be a new strategy for **treating chemotherapy-resistant Ewing sarcoma**. http://www.ncbi.nlm.nih.gov/pubmed/21085683