Chest Wall Tumors

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- benign and malignant etiologies
- *More than half* are malignant

- ***most*** metastases or direct invasion from adjacent tumors of the thorax, mediastinum, or soft tissue.
- Primary chest wall tumors are rare.
- ***Wide surgical resection*** → most effective treatment for the vast majority of chest wall tumors.
- ***Keys to successful management*** include: accurate diagnosis, wide surgical resection, and appropriate reconstruction of large chest wall defects.

Incidence and Location

- The largest institutional experience → Memorial Sloan-Kettering Cancer Center (MSKCC).
- Primary lung and breast cancer made up 163 cases(51%), while 71 patients (22%) had metastatic lesions.

- Primary chest wall tumors, by comparison, are relatively uncommon, representing roughly 5% of all thoracic neoplasms and 1% to 2% of all primary tumors
- The most frequently encountered benign lesions are osteochondroma, chondroma, and fibrous

- Like benign neoplasms, primary malignant tumors generally arise from the soft tissue or the bony and cartilaginous elements of the chest wall.
- In the MSKCC series, soft tissue sarcomas made up 45% of all primary malignant lesions.
- ***most frequent histologies of sarcoma encountered:*** Desmoid tumor, liposarcoma, and rhabdomyosarcoma
- ***most common malignant bone lesions and the most common primary malignant chest wall tumors :*** Chondrosarcoma, Ewing's sarcoma, osteosarcoma
- Chest wall neoplasms, primary and metastatic ➔ ribs, sternum, scapulae, and clavicles.
- ***Primary lesions of the sternum, scapulae, and clavicles are uncommon, but nearly all are malignant***.
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Symptoms and Signs

- -***generally asymptomatic***
- -presenting as ***slowly enlarging masses***.
- -***Generally nonpainful mass***.
- -With continued growth and tumor involvement of surrounding tissue, pain invariably occurs.
- The pain is frequently ***generalized***, and the patient commonly is treated for musculoskeletal or neuropathic etiologies.
- all malignant lesions are likely to become painful, whereas only two-thirds of benign tumors produce pain .
- Cartilaginous and bony tumors more frequently have pain as one of the first symptoms of disease.

Diagnosis

- 1) The initial evaluation : ***complete history and physical examination***.
- history : prior malignancy, exposure to ionizing radiation, or presence of familial conditions, such as Gardner's syndrome or von Recklinghausen's disease.
- 2)Standard conventional ***plain chest radiographs***.
- 3) ***Computed tomography (CT)*** of the chest is the single best radiographic modality.
- 4) ***magnetic resonance imaging (MRI)***

- Compared with CT,MRI can delineate better the extent of muscular invasion, relationship to nearby blood vessels, and/or spinalinvolvement by chest wall lesions. CT, however, is superior in the assessment of calcifications and presence of pulmonary pathology.

- ➔ history, physical examination
- ➔imaging
- ➔ benign or malignant or if the lesion is suspected of being a primary tumor*??????*
- ➔tissue diagnosis by histologic examination ((Biopsy))
- ➔
- *1)* needle biopsy
- *2)* excisional biopsy
- *3)* incisional biopsy.

Needle Biopsy

- minimal soft tissue injury
- ease of performance
- low complication rate in an outpatient setting.
- Fine-needle aspiration alone is best → metastasis from a prior documented malignancy.
- not as good for → primary bony or cartilaginous chest wall tumor(where a larger amount of tissue is required)

Excisional Biopsy

- with minimal margins (1 cm) should be employed for small (<5 cm) primary neoplasms.
- Closure of chest wall ➔ without reconstruction.

- **If** the lesion ➔benign or best treated with non-surgical-therapy (chemotherapy, radiation, or both)➔***no further operation is required.***

- **If** the lesion➔primary malignant neoplasm➔ ***reoperation with radical excision with wide margins and appropriate reconstruction.***

Incisional Biopsy

- 1)FNA nondiagnostic
- 2) for primary lesions >5 cm.

- Skin flaps should not be raised ,and the deep plane of the tumor, in particular the pleural space, should be left intact to prevent dissemination of tumor cells.

- 1-careful preoperative assessment is critical to minimize the risks to the individual patient.
- 2-detailed history and physical examination
- 3-evaluation →**cardiopulmonary comorbidities**, **smoking history**, **performance status**, and **psychosocial support**.
- **pulmonary reserve** is of concern→spirometry, diffusion capacity, and arterial blood gas analysis. Patients with COPD or active pulmonary disease →optimal medical treatment prior to surgery.

Preoperative consultation

- "***medical malignancies***"
  such as Ewing's sarcoma and plasmacytoma➔by medical and radiation oncologists prior to operation is a integral part of their ***multimodality treatment***.

- Depending on the size of the lesion, chest wall reconstruction may warrant the use of myocutaneous flaps; these procedures are best performed in conjunction with ***plastic andreconstructive surgeons***.

- ***neurosurgical consultation*** is often required for tumors encroaching on thev spine

Surgical Management

- In suitable patients, surgical resection for cure is appropriate for primary chest wall neoplasms.
- In select cases of metastatic lesions and locally recurrent breast cancer, surgical extirpation of an isolated chest wall lesion can result in long-term survival benefit.
- Even in instances where the likelihood of cure is low, chest wall resection may be indicated to palliate lesions that are unresponsive to nonsurgical therapies (chemotherapy, radiation) and arecausing local complications, such as wound ulceration, infection, or intractable pain.
- ***The margin of excision*** during definitive resection for chest wall lesions varies somewhat depending on the type of neoplasm. Moreover, what constitutes an adequate margin of excision to minimize the incidence of recurrence and maximize chances for long-term survival for patients with a primary malignant chest wall tumor is a matter of ***some debate***.

Primary Bone Tumors

- uncommon.
- Nearly all were malignant
- sternal tumors > malignant than rib tumors

- The most common benign bone lesions ➔
- Osteochondromas-chondromas-fibrous dysplasia.
- The most common primary malignant bone tumors ➔Chondrosarcoma, Ewing's sarcoma, osteosarcoma

Osteochondroma:

- overall an uncommon tumor

  most common benign bone neoplasm(50% of all benign rib tumors)
- arises metaphyseal region of a rib and develops as a stalked mass with a cartilaginous cap.

- Microscopically, the amount of bony proliferation and thickness of thecartilaginous cap varies.
- Stippled calcification is often present within the tumor, and a rim of calcification can be present at the periphery .
- in childhood and continues to grow until skeletal maturity isreached.
- ***grow inward***, remaining ***asymptomatic***, or ***grow outward***, producing a ***palpable mass***.

Osteochondromas detected in ***children*** after puberty or in adults should be resected.

Before puberty, resection is indicated for the onset of pain or increase in size, especially because malignant degeneration has been reported.

Males are affected 3 time > females.

Complete surgical resection is the treatment ofchoice.

Recurrence has never been reported.

Chondroma

- ***15%*** of all benign rib neoplasms
- most commonly second or third decade of life
- affecting both sexes equally.
- :slowly enlarging, asymptomatic masses originating anteriorly at the costochondral junction.
- radiographic appearance is that of an expansible medullary mass that causes thinning of the cortex.
- Clinically and radiographically, it is not possible to differentiate chondroma fromchondrosarcoma. Histologically, the tumor consists of lobules of hyaline cartilage.
- ***Microscpicaly differentiation between a chondroma and a low-grade chondrosarcoma can be difficult***.
- ***all chondromas must be managed as malignant lesions, with wide excision to prevent local recurrence***.

Fibrous Dysplasia

- Fibrous dysplasia is a benign, cystic lesion characterized by fibrous replacement of the medullary cavity of the rib.It accounts for up to 30% of all benign chest wall tumors and most commonly presents as a solitary mass in thelateral or posterior rib cage. Males and females are equally affected, and multiple lesions can occur in Albright'ssyndrome (multiple bone cysts, skin pigmentation, and precocious sexual maturity in females). These lesions areslow-growing and remain asymptomatic unless they become large enough to cause symptoms through localcompression or pathologic fracture. The radiographic findings are characteristic: a trabeculated, expansile lesionwith a ground-glass center and thinning of the cortex. Microscopic examination reveals bony trabeculation, fibroustissue, and occasionally calcification. Malignant degeneration is unusual, and treatment should be conservative.Resection is curative and is indicated to rule out malignancy and for painful, enlarging lesions.

Eosinophilic Granuloma

- Eosinophilic granuloma is not a neoplasm, but one subset of a spectrum of diseases of the reticuloendothelial systemthat fall under the moniker of histiocytosis X. This also includes Letterer–Siwe disease (infants) andHans–Schüller–Christian disease (children), both of which are characterized by systemic signs and symptoms inaddition to bony lesions. These include fever, malaise, weight loss, lymphadenopathy, and splenomegaly associatedwith leukocytosis, eosinophilia, and anemia. Eosinophilic granuloma is limited to bone involvement and typicallyoccurs in young to middle-aged adults. The radiographic appearance of the bone abnormality is similar for all three:an expansile lesion with periosteal new bone formation and uneven destruction of the cortex, which producesendosteal scalloping. The skull is most commonly involved, but 10% to 20% of patients have rib lesions. Excisionalbiopsy is required for diagnosis and is curative in patients with solitary eosinophilic granuloma. Patients withmultiple lesions should undergo low-dose radiation therapy. In the systemic forms of the disease, the course tendsto be protracted, requiring treatment with chemotherapy and corticosteroids.

Chondrosarcoma:

- ***most common primary chest wall bone neoplasm***
- one-third of all primary malignant bone lesions.
- typically arises in the costochondral arches of rib or sternum in over 60% of all cases.
- frequently in males
- rare <20 years , usually third or fourth decade.
- cause is unknown, although malignant degeneration of benign cartilaginous tumors has been reported.
- association between trauma and chondrosarcoma. ?????!!!!

- Patients develop a slowly enlarging mass that eventually becomes ***painful***.
- ***Radiographically:***
  alobulated mass arising in the medullary portion of the bone with destruction of the cortex and mineralization of thetumor matrix, which produces a mottled type of calcification
- ***Microscopically:***
  findings can range from normal cartilage to obvious malignant changes with plump, atypical, multiple nuclei; these may be more apparent in the periphery of the tumor
- ***Definitive diagnosis:***
- can only be made pathologically, and this often requires abundant tissue in order to distinguish a well-differentiated chondrosarcoma from a chondroma.

Prognostic Factors in Primary Chest Wall Chondrosarcoma

	5-Year survival
No metastasis	79%
Metastasis	27%
Complete resection	69%
Incomplete resection	50%
No resection	20%
No recurrence	88%
Local recurrence	52%
Distant recurrence	37%
Age <50 years	76%
Age >50 years	51%

The treatment of chondrosarcoma

- ***is complete resection***.

- **the factors associated with prolonged survival :**
- lack of metastases, complete resection, lack of recurrence, andage <50 years

- ***Gender, site of primary, size, and grade were not prognostic***.

Ewing's Sarcoma

is a small round-cell sarcoma that occurs primarily in flat bones and the midshaft of long bones

-Primary Ewing's sarcoma of the chest wall represented 15% of all Ewing's sarcomas and 17% of all primary malignant chest wall tumors treated at our institution over a 40-year period.

-Two-thirds of cases occur in patients <20 years ofage

-males are affected twice as often as females.

Almost all patients present with a painful, enlarging massassociated with fever, malaise, leukocytosis, anemia, and an increased erythrocyte sedimentation rate.

The radiographic features can mimic osteogenic sarcoma, osteomyelitis, or other bone tumors. !!!!!!

Mottled destructioncontaining lytic and blastic areas appears, and elevation of the periosteum and multiple layers of subperiosteal newbone formation can cause an onion-skin appearance of the bony surface. Pathologic fractures are

- n the MSKCC experience reported by Burt and coworkers,6 23% of patients had synchronous metastases atpresentation; 71% of those who presented with only local disease developed distant metastases. For this reasonEwing's sarcoma should be considered a “medical tumor,” with the surgeon's initial role being to establish a diagnosis either by incisional or core needle biopsy. Following systemic chemotherapy, the primary site shouldeither be irradiated or resected. With combined-modality treatment, the overall 5- and 10-year survival at ourinstitution was 48%, with a median survival of 57 months. More recent multicenter trials of induction chemotherapyfollowed by resection, radiation, or both demonstrated 5-year event-free survival rates of 62% to 68%.38,39

Osteosarcoma

- Osteosarcoma of the bony thorax is less common than chondrosarcoma, constituting 10% of all primary malignantchest wall tumors in our 40-year experience and 3% of all osteosarcomas. It is, however, a more virulent neoplasmwith a poor prognosis. It generally occurs in teenagers and young adults, with a slight predilection for males. Mostpatients present with a rapidly enlarging, painful mass and elevated levels of serum alkaline phosphatase. Bonedestruction—with indistinct borders that merge into adjacent normal bone—appears on radiographs (Fig. 49-4).Calcification occurs at right angles to the bony cortex, producing a sunburst appearance. Pathologic fractures arerare. The tumor grossly is large, lobulated, and extends through the cortical bone into the adjacent soft tissue.Microscopically, the predominant component may be bony, cartilaginous, or fibrous.In our series of patients, 34% had synchronous metastases; resection was the primary therapy in 82%. Adjuvantchemotherapy was used in 48% of resected patients. Over two-thirds of the patients (68%) developed distantmetastases at some point in their course. The overall 5-year survival was 15%, with a median survival of 12 months.No patients who developed distant metastasis survived 5 years. Approximately half of resected patients receivedadjuvant chemotherapy yet enjoyed no improvement in survival. Our current approach is to give patients inductionchemotherapy followed by wide resection.

Solitary Plasmacytoma

- Solitary plasmacytoma to a rib is uncommon, representing only 6% of all primary chest wall malignancies and 3% ofall plasmacytomas seen in 40 years at MSKCC. Myeloma is most common in the fifth through seventh decades of life,and two-thirds of those afflicted are male. Pain is the most common symptom, often without an associated mass.Most patients are anemic and have an elevated erythrocyte sedimentation rate. Abnormal protein electrophoresis ispresent in 85%, and up to 50% have urinary Bence Jones protein and hypercalcemia. Radiographically, myelomaappears as an osteolytic lesion with cortical thinning. Pathologic fracture is common. Histologically, sheets ofclosely packed, hyperchromatic, and multinuclear cells with abundant cytoplasm and rare mitoses are seen.The majority of our patients (67%) in previous series received primary chemotherapy followed by either resection(one-third) or radiation therapy (two-thirds). 7 The overall 5- and 10-year survival was 38% and 21%, respectively,with a median survival of
  P.67456 months. Notably, 75% of patients with a solitary plasmacytoma of the chest wall developed multiple myeloma. Asin Ewing's sarcoma, the primary role of the surgeon is to establish a diagnosis either by core needle, incisional, orexcisional biopsy. As described in most modern series, the standard primary treatment modality for solitaryplasmacytoma is radiation.8,11,14,19 The major prognostic factor for survival is the development of multiplemyeloma.

Primary Soft Tissue

- TumorsPrimary soft tissue tumors may arise from any cell type of the thorax and may be benign or malignant. Thepredominant benign tumors involving the chest wall include fibromas, lipomas, giant cell tumors, neurogenictumors, vascular tumors (hemangiomas), and connective tissue tumors. Neurogenic tumors involving the bony thorainclude neurilemomas and neurofibromas. Neurofibromas can occur as isolated lesions or in association with vonRecklinghausen's disease (neurofibromatosis).25 Neurilemomas usually occur as solitary tumors that resembleneurofibromas but are not associated with neurofibromatosis. While sarcomatous degeneration may occur inneurofibromas and, to a lesser extent, in neurilemomas, malignant degeneration of benign lesions overall isuncommon, and all can be treated by local excision. Malignant soft tissue lesions and, in particular, soft tissuesarcoma account for approximately 50% of all primary malignant chest wall tumors. Preoperative differentiationbetween the various neoplasms, however, can be difficult. When in doubt, as with bony lesions, wide resection oftumor with surrounding structures is the preferred treatment.

Desmoid

- Desmoid tumors are locally invasive tumors with a propensity to recur following resection. They arise in extremitylocations most commonly (42%–51%). The chest wall has been reported as a site of origin in 10% to 28% of patients. 34 n a past MSKCC series, desmoids represented the most common primary chest wall sarcoma, making up 21% of allhistologic subtypes treated.16 Desmoid tumors affect males and females equally, most frequently betweenadolescence and 40 years of age, and can be associated with Gardner's syndrome. The tumor originates in muscleand fascia, extending along tissue planes and displacing and often encasing surrounding structures, including vessels(Fig. 49-5). Initially asymptomatic, desmoids involving the thoracic inlet can cause paresthesias, hyperesthesia, andmotor weakness, with progressive neural encasement. Histologically, there is a spreading pattern of uniform, well-differentiated fibroblasts and fibrocytes with an abundant intercellular matrix without mitoses or necrosis.Frequently, there are finger-like projections of tumor infiltrating the surrounding tissue well beyond the grossextent.Based on these microscopic findings and because desmoids do not tend to metastasize, some authors consider themto be benign fibromatoses. 15,18 Others, however, consider desmoid tumors to be malignant, low-gradefibrosarcomas because of their aggressive local invasion and propensity to recur, consequently causing morbidity.Desmoid tumors should be treated with wide surgical resection. Death from disease following resection is rare, butlocal recurrence is common. Brodsky and associates4 P.675reviewed our institution's experience, reporting an overall 5-year survival of 93% and a 5-year local recurrence rateof 29% following resection. On univariate analysis, the only factor associated with a higher risk of recurrence wasage >30 years at the time of diagnosis. We have employed intraoperative brachytherapy when wide resection waslimited by the tumor's proximity to vital structures. Recurrence should be treated, when feasible, with repeatresection. Patients with multiply recurrent tumor should be considered for adjuvant radiotherapy. Options fortreatment for patients in whom surgery is not feasible include hormonal therapy, nonsteroidal anti-inflammatorymedication, and chemotherapy.

Soft Tissue Sarcoma

- While soft tissue sarcomas of the chest wall account for one-half of primary malignant chest wall lesions and themajority of metastatic lesions as well, they are still relatively uncommon. Burt7 reported that primary chest wallsarcomas represented only 6% of all soft tissue sarcomas seen over a 40-year period at MSKCC. Men are affectedtwice as often as women.16 The majority of primary chest wall sarcomas occur in adult life; the exception isrhabdomyosarcoma, which is seen most frequently in children and young adults <45 years of age. Most patientspresent with a painless mass, and a wide range of histologic subtypes is seen (Table 49-3).As with soft tissue sarcomas in other sites, primary chest wall sarcomas are generally treated primarily with widesurgical resection. In our series of 149 patients, 140 were treated primarily with resection; 70 underwent resectionat our institution. 16 Local recurrence developed in 59% overall and in 27% of those that had their resection atMSKCC. Neither margin status nor grade of tumor was associated with a higher incidence of local recurrence. Fifty-one percent of patients with high-grade sarcoma developed metastases, compared with only 10% of patients withlow-grade histology. Overall 10-year survival in high-grade sarcomas was 56%, with a significant difference insurvival between high-grade disease (39%) and low-grade disease (82%), patients who developed metastases (19%)versus those who never developed metastases (75%), and pain at presentation (37%) compared with no pain (63%).Because of higher rates of recurrence, metastases, and poorer overall survival in high-grade chest wall sarcoma,most are approached with multimodality therapy. Several authors have published series in which these lesions weretreated with a variety of combined modalities that included induction therapy, surgical resection, and adjuvanttherapy. 33,43 Induction and adjuvant therapies included chemotherapy alone, radiotherapy alone, andchemoradiotherapy. The overall 5-year survival rates approximate 60%. One form of soft tissue sarcoma that issensitive to chemotherapy is rhabdomyosarcoma. This fact, coupled with an otherwise dismal survival with resectionalone (21% 10-year survival), has resulted in current treatment regimens that include induction chemotherapy,complete surgical excision, and adjuvant therapy with radiotherapy, chemotherapy, or both.35

Radiation-Associated Malignant Tumors

- In the last 25 years there have been four series describing postirradiation chest wall sarcomas. The two largestreported rates—of 4.8% and 6% of all primary chest wall sarcomas, respectively—reflect the uncommon nature ofthis problem.37,40 The most common indications for radiation were for breast carcinoma or lymphoma. In the MSKCCexperience, the median latency period between irradiation and development of a primary chest wall sarcoma was 7years (range 2–19), and although a wide P.676range of histologies were observed, osteosarcoma made up over one-half of the 21 lesions.37 The median dose ofradiation was 4,140 cGy (range 1,250–9,500), with most patients receiving external beam radiation. The precisemechanism of carcinogenesis for postirradiation sarcomas is not known. The results of treatment of lesions arising ina previously irradiated field were comparable to their de novo counterparts, with similar survival rates followingwide surgical resection. Thus, these patients should be treated in a manner commensurate with that typicallyemployed for the de novo pathologic tumor type.

- Tumors of the Sternum, Scapula, and ClaviclePrimary chest wall tumors, benign and malignant, and metastatic lesions can frequently involve the manubrium andsternum. Benign lesions are typically chondromas, bone cysts, or hemangiomas. Metastatic tumors usually originatefrom carcinomas of the breast, thyroid, or kidney. In the MSKCC series, the sternum was the site of origin in 14% ofbony and cartilaginous tumors overall. 5 Chondrosarcoma is the most common histologic type, followed closely byosteosarcoma, plasmacytoma, and lymphoma. Complete surgical resection via partial (<50%), subtotal, or totalsternectomy with repair of defects employing rigid prostheses is a safe and effective treatment.21,26,31 Overallsurvival following complete surgical resection is related to the tumor histologic type and grade.The scapula is a common site for primary bone tumors but an infrequent site for metastatic lesions. As Burt andcolleagues5 noted, the scapula was the site of origin for malignant bony and cartilaginous lesions in 31% overall. Themost common histologies include chondrosarcoma, Ewing's sarcoma, and osteosarcoma, all of which occur withequal frequency. Wide surgical resection is the treatment of choice. In contrast, with the exception ofplasmacytoma, the clavicle is an uncommon site for primary malignant bony tumors, accounting for the site of originin only 8% of cases. In fact, the clavicle is more likely to be a site of metastatic disease rather than primary tumor.Most clavicular lesions are malignant and should be treated with primary radiotherapy or complete surgicalresection. Reconstruction following resection is generally unnecessary.

- Metastatic Disease and Recurrent Breast CarcinomaThe role of surgical resection for metastatic disease to the chest wall or locally recurrent breast carcinoma iscontroversial, because the former reflects disseminated disease and the latter portends it. As previously mentioned,metastatic disease accounts for 20% to 30% of all chest wall neoplasms and can occur within the bony thorax or thesoft tissues surrounding the ribs, sternum, scapulae, or clavicle. The majority cannot be cured by surgicalextirpation, but resection can be considered in certain cases for cure and in others to palliate pain or ulceration andinfection of the overlying skin. Anderson and coauthors1 proposed the following criteria for curative resection: (a)the chest wall is the only site of disease, (b) locoregional disease is controlled, and (c) complete resection withnegative margins is possible. Martini and coworkers26 reported a 20% 5-year survival for chest wall metastasectomy,while 41% of patients were alive at a median follow-up of 31.5 months in the series by Pairolero and Arnold.31Up to 10% to 12% of stage II breast carcinomas recur locally after mastectomy, sometimes with chest wallinvolvement.
  P.677As demonstrated by Valagussa and coauthors, 41 the majority recur within the first 3 years, and up to 80% of patientswith locoregional recurrence will develop distant disease. Disease isolated to the chest wall and a disease-freeinterval from mastectomy to recurrence longer than 2 years correlates with survival following surgical resection oflocal chest wall recurrence. McCormack and associates30 reported a 50% 5-year disease-free survival in 35 patientswho underwent chest wall resection. As reviewed by Incarbone and Pastorino, 22 a number of small series show 5-year survival rates that range from 35% to 58% following curative resection. Aggressive treatment of local failure, inaddition to the potential curative benefit, results in palliation of pain, removes a potentially unsightly wound, andachieves optimal local control of disease.

Conclusions

- Successful management depends on timely diagnosis, careful patient evaluation, and aggressive surgical resection, with adequate chest wall reconstruction where appropriate.

- With modern surgical and reconstructive techniques, this procedure can beperformed in a single operation with minimal pulmonary compromise and low operative morbidity and mortality.
- in the majority of primary chest wall tumors, complete and wide surgical resection can lead to potentialdisease-free and overall survival. In the case of certain histologic subtypes, multimodality therapy combiningchemotherapy, surgical resection, and radiotherapy in both induction and adjuvant settings is appropriate. Surgicalmanagement of metastatic tumor and recurrent breast carcinoma is often necessary for palliation and in isolatedcases can lead to prolonged survival.