Prognosis

Conventional Imaging

Even dynamic contrast-enhanced MRI after preoperative chemotherapy cannot yet give an accurate prediction of lasting tumor remission (Reddick et al. 2001).

FDG PET

With osteosarcoma, the initial FDG uptake of the primary tumor is a prognostic indicator (Franzius et al. 2002b). Intense glucose hypermetabolism is associated with a reduction of overall and disease-free survival. This is consistent with the result of a large study in more than 200 adults and children with bone and soft-tissue sarcomas (Eary et al. 2002).

A prospective study of Ewing tumors demonstrated the prognostic value of glucose metabolism following neoadjuvant chemotherapy. In this study, too, high FDG uptake correlated with a shorter period of disease-free survival (Hawkins et al. 2005).

PET-CT

No studies have yet been published on the prognostic capabilities of PET-CT in patients with primary bone tumors.

Soft-Tissue Sarcomas

Basic Considerations

Soft-tissue sarcomas in children are a heterogeneous group of malignant tumors that originate in the soft tissues and have a predominantly mesenchymal origin. The most common histologic entities in children and adolescents are rhabdomyosarcoma (embryonal and alveolar, 61%), extraosseous Ewing sarcoma and peripheral neuroectodermal tumor (PNET, 8%), synovial sarcoma, neurofibrosarcoma, fibrosarcoma, and leiomyosarcoma (Kaatsch and Spix 2006). There is no uniform system for the staging and risk grouping of soft-tissue sarcomas. Pretreatment biopsy is always necessary. The diagnosis of soft-tissue sarcoma is based on morphologic and immunohistochemical criteria. The 10-year survival rate is 60% (Kaatsch and Spix 2006).

Conventional Imaging

Ultrasonography is often the initial imaging study for soft-tissue sarcomas, depending on lesion location and accessibility. Other sectional imaging studies are needed for evaluation of tumor extent, however. MRI provides significantly better soft-tissue contrast, does not involve ionizing radiation exposure, and is therefore preferred over other modalities. Pretreatment staging should also include chest radiographs in two planes, thoracic CT, cranial MRI, and abdominal ultrasonography or MRI (Gadner et al. 2006; AWMF 2007).

PET and PET-CT

No clinical studies have yet been published on the evaluation of pediatric soft-tissue sarcomas with FDG PET and PET-CT. In one series in which PET-CT was used in three patients with rhabdomyosarcoma it was concluded that slight FDG uptake in lymph nodes requires biopsy confirmation as it may indicate either metastasis or nonspecific activity (Ben Arush et al. 2006). In another series of four patients with rhabdomyosarcoma it was concluded that FDG PET is useful for evaluating response to therapy (Peng et al. 2006).

The great majority of soft-tissue sarcomas in children show intense glucose hypermetabolism (**Figs. 2.82, 2.83, 2.84, 2.85, 2.86, 2.87**). It is reasonable to conclude, then, that FDG PET is clinically useful for staging and for evaluating treatment response. Two larger studies of soft-tissue sarcomas in adults and some children have shown that FDG PET is not useful for the exclusion of pulmonary metastases, analogous to the results with bone sarcomas (Lucas et al. 1998; Iagaru et al. 2006). In cases where PET is proposed for staging, it would definitely be advantageous to obtain simultaneous CT images on a PET-CT scanner (**Fig. 2.88**)

Neuroblastoma

Basic Considerations

Neuroblastoma is the most common solid extracranial malignant tumor in children. Its incidence is 1.1:100 000 children under 15 years of age (Kaatsch and Spix 2006). As an embryonal tumor, neuroblastoma is most frequently diagnosed in infants and small children. It originates from cells of the neural crest and thus occurs predominantly in the sympathetic trunk, paraganglia, and adrenal medulla. Tumors of the sympathetic nervous system show increased catecholamine production in more than 80% of patients.

Staging is based on the International Neuroblastoma Staging System (INSS). Stage IV, characterized by the presence of distant metastasis, has a particularly grave prognosis. Molecular genetic characteristics also have prognostic significance (e.g., N-*myc* amplification). The 10-year survival rate is 53% for all patients but varies

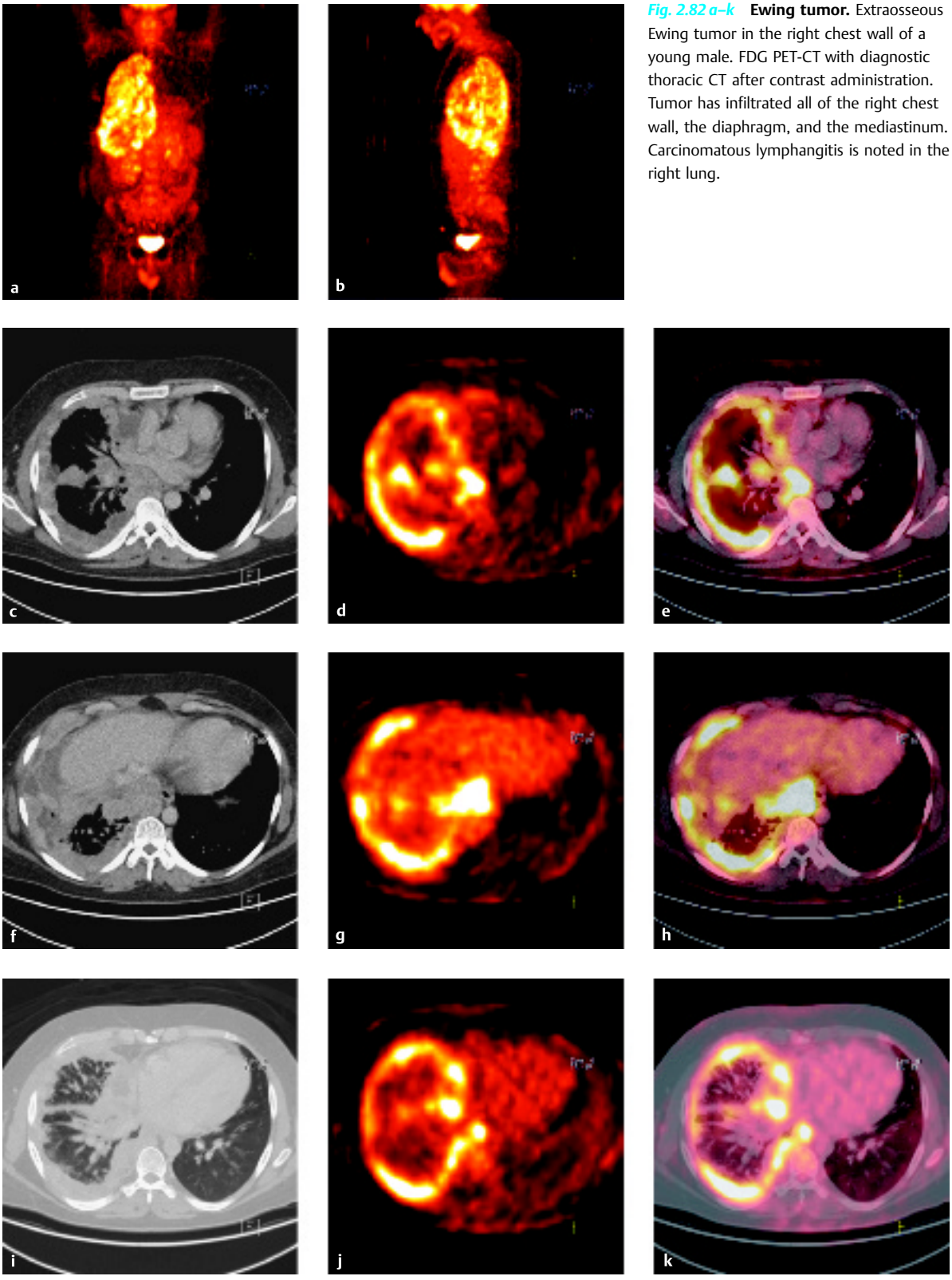

Fig. 2.82 a–k **Ewing tumor.** Extraosseous Ewing tumor in the right chest wall of a young male. FDG PET-CT with diagnostic thoracic CT after contrast administration. Tumor has infiltrated all of the right chest wall, the diaphragm, and the mediastinum. Carcinomatous lymphangitis is noted in the right lung.

a FDG PET, coronal whole-body MIP.
b FDG PET, sagittal whole-body MIP.
c–h Axial scans of the chest at two different levels (**c, f** CT with a soft-tissue window; **d, g** FDG PET, **e, h** PET-CT).
i–k Axial scan through the chest (**i** CT with a lung window, **j** FDG PET, **k** PET-CT).

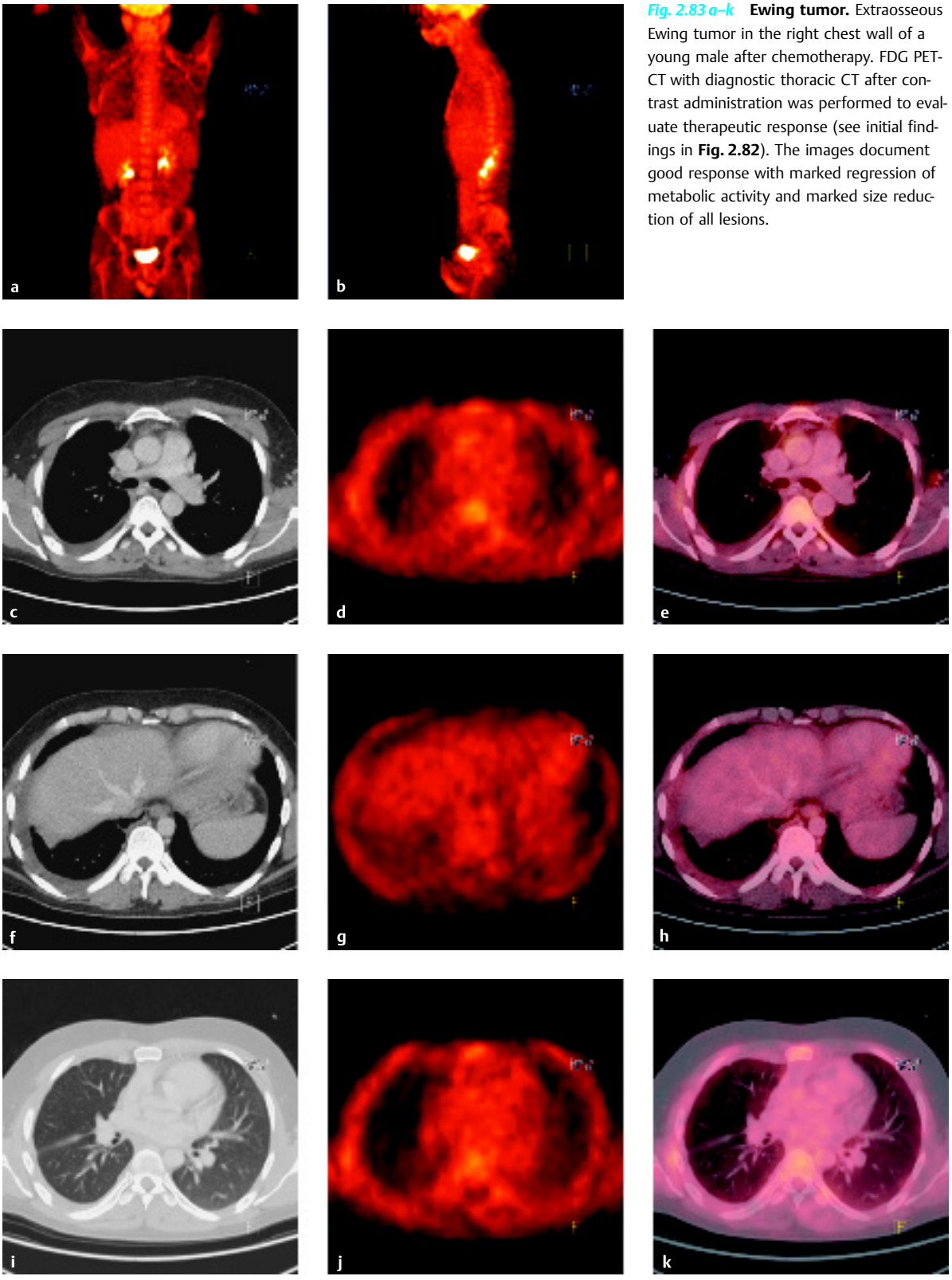

Fig. 2.83 a–k **Ewing tumor.** Extraosseous Ewing tumor in the right chest wall of a young male after chemotherapy. FDG PET-CT with diagnostic thoracic CT after contrast administration was performed to evaluate therapeutic response (see initial findings in **Fig. 2.82**). The images document good response with marked regression of metabolic activity and marked size reduction of all lesions.

a FDG PET, coronal whole-body MIP.
b FDG PET, sagittal whole-body MIP.
c–h Axial scans of the chest at two different levels (**c, f** CT with a soft-tissue window; **d, g** FDG PET; **e, h** PET-CT).
i–k Axial scan through the chest (**i** CT with a lung window, **j** FDG-PET, **k** PET-CT).