Central nervous system tumors constitute the largest group of solid neoplasms during childhood and are second only to leukemia in their overall frequency in children. Each year, approximately 1500 to 2000 children develop brain tumors in the United States; at least 50% of these tumors are gliomas. These lesions constitute a diverse group in terms of their histopathological appearance, molecular biological characteristics, and response to therapy. This special edition of the Journal of Neuro-Oncology is devoted towards recent advances in the diagnosis and treatment of these often challenging tumors.

In the first chapter, Dr. Raffel discusses the molecular and cytogenetic techniques that have been used to characterize brain tumors, and examines the features of pediatric gliomas that distinguish these lesions from related neoplasms in adults. In contrast to the results in adult high-grade astrocytomas, childhood malignant gliomas less commonly exhibit mutations of the p53 gene, amplifications or rearrangements of the epidermal growth factor gene, or deletions of chromosome 10. Taken together, these observations imply that the pathways leading to the development of malignant astrocytomas in children may differ significantly from those involved in adults, which may in part account for the fact that these lesions seem to carry a somewhat better prognosis in children. Interestingly, childhood brainstem gliomas, which are similar to adult malignant gliomas with regard to their dismal prognosis, exhibit many of the same chromosomal abnormalities as the adult tumors. Dr. Raffel also presents some intriguing data that suggest a possible relationship between mutations in the neurofibromin gene and the development of pilocytic astrocytoma, which is the most common central nervous system tumor in patients with neurofibromatosis 1.

The next several chapters deal specifically with the management of gliomas in different locations within the brain. Dr. Berger reviews the role of various technical adjuncts in the management of cerebral hemispheris low-grade gliomas. In particular, it is now possible to appropriately target the operative approach to a deep subcortical lesion or a superficial lesion that is located within ‘eloquent’ cortex using a combination of functional studies and stereotactic localization. These techniques enable the surgeon to minimize injury to critical areas of the brain while proceeding with aggressive removal of lesions that previously might have been considered to be unresectable. This chapter also contrasts the long-term outcome of low-grade gliomas in children vs. adults. In contrast to adult astrocytomas, childhood tumors rarely progress histologically to more malignant lesions and long-term disease control is often achieved after aggressive resection.

Until recently, thalamic astrocytomas were considered to be largely unresectable based on the fact that conventional surgical approaches carried a prohibitive risk of permanent neurological morbidity. Drs. Souweidane and Hoffman discuss recent technical adjuncts that have facilitated aggressive resection of these lesions, review the indications for surgical intervention, and present guidelines for
postoperative management. With the implementation of computer-assisted stereotactic approaches, perioperative morbidity and mortality have dropped precipitously and near complete resection has become an attainable goal in many children with low-grade lesions. In such cases, adjuvant therapy can often be deferred. In contrast, for patients with malignant glioma, tumor debulking, if indicated to relieve local mass effect, must be followed by adjuvant radiotherapy and/or chemotherapy. Even with aggressive multimodality therapy, the prognosis for long-term survival remains poor.

Chiasmatic-hypothalamic gliomas constitute a diverse group of tumors in terms of their biological behavior. Although some lesions remain quiescent for years without therapy, others progress rapidly despite multimodality treatment and ultimately prove fatal. The majority of these lesions are not amenable to radical surgical debulking because of their diffuse involvement of the optic apparatus and hypothalamus. Although radiotherapy has long been a cornerstone in the treatment of such unresectable tumors and is effective in producing long-term disease control in the majority of patients, late cognitive and endocrine impairment are a major concern, particularly in children younger than five. Accordingly, chemotherapy has come to play an increasing role in the management of these tumors in young patients. Drs. Garvey and Packer discuss the relative roles of surgery, radiotherapy, and chemotherapy in contributing to an integrated approach to the treatment of these lesions.

Supratentorial malignant gliomas remain a challenging management problem. Although these tumors have previously been treated with aggressive resection followed by postoperative radiotherapy, the vast majority of affected children died of disease progression. During the last decade, several studies have demonstrated a clear benefit of chemotherapy in children with glioblastoma multiforme. However, most patients have still succumbed to progressive tumor growth. Accordingly, recent studies have been examining the effect of more intensive treatment regimens in the hope of improving the outcome of children with these tumors. Drs. Lyden, Mason, and Finlay review recent advances in the chemotherapeutic treatment of these neoplasms and present the preliminary results that have been achieved with regimens that couple intensive chemotherapy with autologous bone marrow reconstitution.

The management of brainstem gliomas has been refined substantially since the advent of magnetic resonance imaging and the recognition that different types of lesions exhibit characteristic imaging features that correlate with biological behavior. Whereas certain subgroups of tumors are focal, histologically benign, and amenable to radical resection, other lesions infiltrate the brainstem diffusely, are highly malignant biologically, and are best managed nonsurgically. Drs. Constantini and Epstein review the indications and technical considerations for the surgical management of benign brainstem gliomas and provide a thorough discussion of the potential risks of surgery versus other therapeutic modalities for the management of these tumors. These lesions include dorsally exophytic tumors arising from the fourth ventricular floor, cystic and solid tumors arising focally within the brainstem, and gliomas of the cervicomedullary junction. In skilled hands, appropriately selected lesions can often be radically resected with acceptable morbidity and, in many cases, long-term survival is possible without the need for adjuvant therapy.

In contrast to the generally favorable results that have been achieved with focal brainstem gliomas, diffuse intrinsic tumors constitute a much more discouraging group. These lesions are not amenable to resection and, in view of their characteristic imaging features, rarely require biopsy. Thus, their management relies almost exclusively on radiotherapy and chemotherapy. Unfortunately, even with high-dose radiotherapy and intensive chemotherapy, the long-term prognosis is poor with median survivals of less than one year in most studies. Drs. Jennings, Freeman, and Murray review the results of recent trials for patients with these tumors and discuss the role and rationale of innovative radiotherapeutic and chemotherapeutic strategies for the management of these lesions.

Cerebellar astrocytomas constitute a final group of childhood gliomas that is widely known for its generally favorable prognosis. However, a number of factors appear to have an impact on outcome. In