Optic Pathway Tumors in Children

Tu mors of the optic path ways are sub-divided in this re view into those that arise in one or both optic nerves at the chiasm (optic nerve tu mors); those that arise within the chiasm and do not extend significantly into the hypothalamus (optic chiasmatic tu mors) and the large exophytic tu mors that in volve both the optic chiasm and the hypothalamus to a lesser or greater degree (optic chiasmatic/hypothalamic tu mors). The man age ment of optic chiasmatic gliomas is contro versial, partly related to fail ure to sep a rate out chiasmatic tu mors from the chiasmatic/hypothalamic tu mors. The optic nerve tu mors are re viewed briefly, since they rarely ex tend intracranially. Chiasmatic tumors tend to be associated with NF1 and to behave almost like hamartomas. Close ob ser vation is usually the most ap propri ate man age ment. On the other hand, chiasmatic/hypothalamic tu mors grow like typ i cal neoplasms. The tu mors are al most uni formly low grade astrocytomas, but growth rates may be rapid, es pe cially in in fants. Modern man age ment has trended away from rad i cal sur gi cal re sec tion, which has sig nif i cant mor bid ity, to che mo ther apy as the first line of treat ment. In this re view, the clin i cal pre sen ta tion and man age ment of dif fer ent types of optic path way tu mors are dis cussed.

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Op tic path way tu mors are re latively un com mon, com prising only 1 per cent of brain tu mors at all ages. The ma jor ity of these tu mors pres ent in the pe di at ric pop u la tion and optic path way tu mors make up be tween 4 and 6 per cent of all pe di at ric brain tu mors.1 In the adult pop u la tion optic path way tu mors are usu ally anaplastic astrocytomas or glioblastomas with a poor prog no sis. In the child, these tu mors are al most in vari ably low grade astrocytomas, the ma jor ity of which are pilocytic astrocytomas (WHO Grade 1).

There is a well rec ognized as so ci a tion be tween optic path way gliomas and NF1. Ap prox i mate ly 20 per cent of all pa tients di ag nosed with an optic path way glioma have NF1 and among pa tients with NF1 be tween 1.5 and 15 per cent will de velop an optic path way glioma at some time, with symp toms re lat ing to this oc cur ring in half the pa tients.2 Op tic path way gliomas are al most al ways low grade astrocytomas, ei ther pilocytic or dif fuse. De spite the uni formly low grade astrocytomas, the rate of growth with or without therapeutic intervention is unpre dictable.3 As a result, the man age ment of optic path way gliomas is contro versial. Some authors have stated that these neoplasms behave like hamartomas and thus treat ment should be con servative,4 while most oth ers have found the tu mor be hav ior to be more sin ister.5-7

There ap pears to be a re la tion ship be tween the age, pre sen ta tion and the rate of growth of the tu mor, with tu mors oc cur ring un der the age of 5 and par tic u larly in in fants tend ing to grow more rap idly than those tu mors that pres ent later on in child hood.8 As men tioned above, tu mors that pres ent in adult life are al most al ways ma l ignant astrocytomas or glioblastomas, which grow rapidly and are le thal. The nat ur al his tory of pe di at ric low grade optic path way astrocytomas is fu rther con fused by some re ports of spontaneous re mis sion of tu mors. The in flu ence of as so ci ated NF1 on the tumor bi ology has not been defined, but in general the optic path way astrocytomas as so ci ated with NF1 tend to be slower grow ing than those that oc cur in iso la tion.12-14 Tu mors of the optic path ways may be sub-divided, based on anatomic cal lo-

Key Words
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- optic chiasm
- hypothalamus
- optic nerve

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The management of the optic nerve glioma depends on the primary site of origin. This phenomenon has been observed in a significant number of patients, which tends to be associated with NF1. In the majority of reported studies about optic pathway gliomas, we have separated these tumors, for this review, from other tumors of the optic pathway in children, as those tumors, which arise in the region of the optic chiasm and do not extend significantly anterior to the chiasm (optic nerve tumors). Those that arise within the chiasm and do not extend significantly posteriorly to the optic chiasm, rather than the larger tumors growing into the hypothalamic region, tend to be more like a hamartoma than a neoplasm and surgical intervention is often required. As is the case for the asymptomatic tumors, the presence of associated NF1 tends to be a positive prognosticator. These optic gliomas as so ciated with NF1 tend to be more like a hamartoma than a neoplasms and surgically in the vertebrae is often the most required.

**Tu mors of the op tic chiasm**

Optic chiasmatic tumors are defined for this review as those tumors, which arise in the region of the optic chiasm with a relatively small local extension beyond the confines of the enlarged chiasm. Whereas these tumors may be associated with tumor involvement anteriorly into the optic nerves or posteriorly into the optic tracts, we have separated these tumors, for this review, from those tumors, which have a large local extension, typically into the region of the hypothalamus. Like the optic nerve tumors, these tumors of the optic pathway in children, these tumors almost invariably are pilocytic astrocytomas. In our experience, it is these optic chiasmatic tumors, rather than the larger tumors growing into the hypothalamus, that tend to be associated with NF1. In the situation, observation is appropriate. There is controversy about how often follow up MR scans should be performed. In our opinion, a re peated scan in one year is reasonable to confirm that there has been no growth of the tumor and certainly if the patient becomes symptomatic with decreasing vision, a repeated scan is indicated. Whether additional scans, for example on an annual basis, are appropriate in an asymptomatic child with known NF1 is disputed. Of these children are young and require sedation and/or general anesthetic to get a MR scan and usually the lesions remain static for many years. In this situation we have tended to cut down on the frequency of MR scans to perhaps once every five years if the patient remains asymptomatic.

In the situation where the patient is asymptomatic at the time of presentation, the management depends on how much vision is preserved in the affected eye. If the vision is useless in the eye, and there is a large tumor causing proptosis, it is reasonable to resect the tumor completely to improve the cosmetic appearance. If useful vision is still preserved, observation may be more appropriate, with consideration of surgical resection only when useful vision has been lost. As is the case for the asymptomatic tumors, the presence of associated NF1 tends to be a positive prognosticator. These optic gliomas associated with NF1 tend to be more like a hamartoma than a neoplasms and surgically in the vertebrae is often the most required.

**Tu mors of the op tic nerve**

Astrocytomas confined to one or both optic nerves are relatively uncommon in pediatric neurosurgical practice. Approximately ten per cent of all optic pathway tumors involve both optic nerves. In the latter situation there is usually a child with NF1. In many centers these optic nerve tumors are managed by orbital or ophthalmologists rather than by neurosurgeons. This group of patients will be reviewed only briefly at this time.

Children with a tumor confined to the optic nerve typically present with visual loss, proptosis related to the bulk of the tumor within the orbit, and either optic atrophy or papilledema. Occasionally the tumors will extend posteriorly along the involved optic nerve to the region of the chiasm and it is in these cases that the pediatric neurosurgeon is often asked for an opinion.

The diagnostic test of choice is an MRI which will usually show the fusiform tumor within the orbit, displacing the globe forward. The relation of the tumor posteriorly to the optic chiasm can usually be readily identified with thin cuts on MRI (Fig. 1).

The management of the optic nerve glioma depends on the presenting features and the presence of NF1. In many children unilateral or bilateral optic nerve gliomas are identified as part of a screening MRI that may be done after a diagnosis of NF1 has been made. These tumors may be asymptomatic. In that situation, these tumors may be asymptomatic. In that situation, the presence of associated NF1 tends to be a positive prognosticator. These optic gliomas associated with NF1 tend to be more like a hamartoma than a neoplasms and surgically in the vertebrae is often the most required.

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a review of 14 such cases at B.C. Children’s Hospital, ten had associated NF1. This was in contrast to the optic chiasmatic/hypothalamic tumors, which will be discussed in the next section, where there was no association with NF1 in our experience.

Optic chiasmatic tumors present with visual loss, typically affecting the vision in both eyes, with major visual field abnormalities. There is usually bilateral optic atrophy at the time of presentation, more marked on one side than the other. Unlike the optic nerve tumors, there is no proptosis present, since there is no intraorbital component.

The diagnosis is best made with MRI, which will often show the tumor with clear involvement of thickened posterior optic nerves and the optic chiasm (Fig. 2). In children with NF1 a frequent finding is signal change on MRI along the optic tracts. Whether this represents tumor or not is not always clear, but when this type of signal change is present it is almost pathognomonic for an optic glioma in the region of the chiasm and obviates the need for a tissue diagnosis. It should be noted that recent reports have indicated that signal changes along the optic tracts may occur in association with other tumors, such as craniopharyngioma and germ cell tumors. In these cases the signal abnormality is thought to represent edema and the optic tract abnormality subsides when the tumor bulk is decreased.

In those cases where there is no signal change along the optic tracts, and where it is not clear that the lesion is confined to the optic

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**Fig. 1.** (A) Sagittal T1 MRI showing intraorbital tumor (arrow). (B) Sagittal T1 MRI showing intracranial component of tumor (arrow). (C) Axial and (D) coronal T1 MRI with gadolinium, showing intracranial tumor extending to just anterior to chiasm.
path ways in the region of the chiasm, one must consider other suprasellar lesions in the differential diagnosis. These include germinoma, sarcoidosis, histiocytosis, craniopharyngioma and the rare childhood pituitary adenoma. Chiasmatic gliomas are usually diffusely contrast enhancing lesions and only occasionally will have a cyst associated with them. Calcification tends not to be present, which is a differentiating feature from craniopharyngioma. Investigations are typically carried out to identify any endocrine dysfunction related to the anterior or posterior pituitary. The finding of diabetes insipidus at presentation in a relatively small suprasellar tumor is strongly against the diagnosis of an optic chiasmatic astrocytoma and much more in favor of the diagnosis of germinoma. In situations where the radiographic and/or clinical findings are not definitive for an optic chiasmatic intrinsic glioma, a tissue diagnosis is appropriate. Because of the small size of the tumor and proximity to the optic chiasm, we have generally obtained this tissue diagnosis by an open procedure rather
than stereotactically. During the biopsy procedure it is
im portant to biopsy from within the lesion itself and not
only on the sur face of the lesion, since gliosis adja cent to
a tumor, such as ger minoma, can be interpreted by the
pathologist as a low grade astrocytoma.

Except for obtaining the tissue diagnosis in situa
tions that are not clear from the MRI, there is no role for a
sur gical intervention in gliomas that are con fined to the
region of the optic chiasm. The chiasmatic tumors are
small, they usu ally do not present be cause of mass ef fect,
so that there no neural struc tures that re quire de com pres-
sion. The appro pri ate man age ment is ob serv ation. In our
experience, out of 14 patients with chiasmatic gliomas
13 were observed over many years and none pro-
gressed.14 The other patient was treated with radia-
ther apy, which we would no longer recom mend. Should
the tu mor pro gress both rad io log i cally and clin i cally, ad-
ditional treat ment is con sid ered. One has to be cau tious
about the diagnosis of progression. It is not unusual to
see changes in the sig nal along the op tic tracts from one
scan to an other in chil dren with an op tic chiasmatic tu-
more and NF1, and these changes may not be as so ci ated
with changes in visual acuity or visual fields. Of ten the
radiographic changes appear to improve, only to get
worse on the fol low ing scan, or vice versa. En hance ment
pat terns may change, with more en hance ment in one area
and less en hance ment in an other area, and the ex tent of the
lesion may fluc tu ate. Rec og nizing that such changes in the
radiographic pat terns do not neces sarily in di cate pro gres-
sion or re gres sion of the tu mor, it is im por tant to con sid er
the clin ic al sit u ation, par tic u larly the vi sual func tion to -
gether with any ra dio graphic changes be fore com ing to a
con clu sion about re gres sion or growth of tu mor.

It is un usual that a tu mor will ac tu ally pro gress de fi-
nitely to the point that one would consider addi tional
 treatment. In these situa tions, chemother apy is the first
approach recom mend ed, with radiation therapy as the
last op tion. These mo dal i ties of treat ment are dis cus sed
more fully in the next section under manage ment of
chiasmatic/hy po thalamic tu mors.

Optic chiasmatic/hy po thalamic tu mors

Optic chiasmatic/hypothalamic tumors are defined
for this review as those tumors involving the optic
chiasm which are exophytic and ex tend sig nificantly into
the region of the hypo thalamus. Un like the “chiasmatic
tumors” which are con fined to the region of the chiasm
and are small, the op tic chiasmatic/hypo thalamic tu mors
by our def inition are larger. These tu mors of ten pres ent
at a mas sive size, with tu mor ex tend ing su per i orly to fill
the third ven tri cle and ob struct the re gion of the fo ram
men of Monro.

Presenting features include visual loss, signs and
symptoms of in creased intracranial pres sure, en do crine
dysfunction and the diencephalic syn drome. The visual
loss is of ten se vere when first noted. Of ten the child will
be blind in one eye at the time of pre sen ta tion. In in fats,
nyst agmus will of ten be an in di ca tor of se vere visual loss
or even total blind ness. Optic atrophy is usu ally pres ent,
and even in cases with other ev idence of in creased in tra-
cranial pres sure, pap il lede ma is un usual. Visual loss
 tends to be asym met ric, and a pos i tive Marcus Gunn phe-
nomenon (swing ing light test) is usu ally pres ent and may
be an im por tant clue in di ag nosing the vi sual loss in the
infant. In creased intracranial pressure results from a
com bi na tion of a very large tu mor and ob struc tive hy dro-
cephalus. In in fats, the fontanel may bulge, the su tures
may be splayed and the head may be dis pro por tion ately
enlarged. Older children present in the more clas si cal
way with head ache and vom iting. With a large le sion in
the re gion of the hy po thala mus it is not un ex pected that
there might be endocrine find ings at present ation. The
most com mon of these is failure to thrive and dience-
phalic syn drome. In this situa tion the pa tient of ten looks
quite ema ci ated and has been in ves ti gated for some time
for other causes of fail ure to thrive. Pre co cious pu berty
has been re ported, par tic u larly in chil dren be tween 1 and
5 years of age. It is in ter est ing that di a be tes insipidus at
the time of pre sen ta tion is un usual. The pre sent ing symp-
toms and signs differ, de pend ing on the age at pre sen ta-
tion (Table 1).

The man age ment of chil dren with op tic chiasmatic/
hypothalamic gliomas involves a num ber of con sid er-
ations. These in clude the patho logic di ag no sis, the man-
age ment of hydrocephalus if pres ent and the manage-
ment of the tumor mass itself. The investiga tion of
choice is a MRI, although a CT scan can be useful in
cases where there is a cys tic and solid le sion, which may

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mimic a craniopharyngioma. In these cases the calcification of a craniopharyngioma will be seen better on a CT scan than on the MRI. The differential diagnosis of the suprasellar mass includes germinoma, teratoma, other germ cell tumors and pituitary adenoma. The optic chiasmatic/hypothalamic gliomas tend to be diffusely enhancing tumors, which cannot be separated from the optic chiasm anatomically on the MRI. The pattern of growth is usually characteristic, with much of the extension being superior and posterior into the region of the third ventricle and hypothalamus (Fig. 3). The tumor does not typically extend into the sella turcica, which is usually not enlarged. This distinction distinguishes the tumor from a pituitary adenoma. The optic chiasmatic/hypothalamic tumors are usually solid, but cystic varieties do occur, in which case the diagnosis of craniopharyngioma might be considered. Germinomas or non-germinomatous germ cell tumors are usually solid, but cystic varieties do occur, in which case the diagnosis of craniopharyngioma might be considered. Germinomas or non-germinomatous germ cell tumors are usually solid, but cystic varieties do occur, in which case the diagnosis of craniopharyngioma might be considered. Germinomas or non-germinomatous germ cell tumors are usually solid, but cystic varieties do occur, in which case the diagnosis of craniopharyngioma might be considered. Germinomas or non-germinomatous germ cell tumors are usually solid, but cystic varieties do occur, in which case the diagnosis of craniopharyngioma might be considered. Germinomas or non-germinomatous germ cell tumors are usually solid, but cystic varieties do occur, in which case the diagnosis of craniopharyngioma might be considered. Germinomas or non-germinomatous germ cell tumors are usually solid, but cystic varieties do occur, in which case the diagnosis of craniopharyngioma might be considered.

If signal change is present along the optic tracts, posterior to the region of the tumor, and particularly if the patient is known to have NF1, one might be confident that the tissue diagnosis of craniopharyngioma is the most appropriate. Germinomas or non-germinomatous germ cell tumors are usually solid, but cystic varieties do occur, in which case the diagnosis of craniopharyngioma might be considered.

Table 1. Presenting symptoms and signs of chiasmatic hypothalamic tumors

<table>
<thead>
<tr>
<th>Infants</th>
<th>Children 1–5 years</th>
<th>Children more than 5 years</th>
</tr>
</thead>
<tbody>
<tr>
<td>Macrocephaly</td>
<td>Headache/vomiting</td>
<td>Gradual visual loss</td>
</tr>
<tr>
<td>Failure to thrive</td>
<td>Mild/severe visual loss</td>
<td></td>
</tr>
<tr>
<td>Severe visual loss</td>
<td>Precocious puberty</td>
<td></td>
</tr>
</tbody>
</table>

If the diagnosis is not clear based on the imaging studies and serum markers for beta-HCG and alphafetoprotein, one has to consider obtaining the tissue diagnosis. In large tumors with associated hydrocephalus, endoscopic biopsy may be most appropriate, in that one might be able to achieve a biopsy of the tumor, which presents at the foramen Monro quite readily and at the same time fenestrate the septum pellucidum. These children typically will require a ventriculoperitoneal shunt, and by fenestrating the septum, one can avoid the placement of two separate ventricular catheters in the presence of a blocked foramen Monro of Monro. When the ventricles are not markedly enlarged, but the tumors still extend superiorly into the third ventricle and hypothalamus sytem. In this situation, fenestration of the septum pellucidum is not necessary. Alternatives to endoscopic biopsy in cludes stereotactic biopsy, either with or without a frame, or open biopsy via craniotomy. The latter may be the most appropriate approach if one has decided to resect part of the tumor, as will be dis cussed below.

In those patients who present with obstructive hydrocephalus, management involves a ventriculoperitoneal shunt. Be cause the foramen Monro is blocked in these situations, separate ventricular catheters have to be placed into either lateral ventricle unless the septum pellucidum has been fenestrated prior to placement of the shunt. One of the problems that has been associated with ventriculoperitoneal shunts in management of hydrocephalus associated with optic chiasmatic/hypothalamic tumors is the later development of CSF ascites. CSF ascites is a very unusual complication of a ventriculoperitoneal shunt but is disproportionately found in patients with optic chiasmatic/hypothalamic tumors.

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Unlike the optic chiasmatic tumors, research has not focused on whether optic chiasmatic/hypothalamic tumors tend to grow, sometimes rapidly, and usually behave like a neoplasm rather than a hamartoma. The growth rate ap pears to be parenteral rapid in infants and youn ger children. The options available for management of the tumor itself include surgical resection, chemotherapy and radiotherapy and each will be dis cussed in turn.
Surgical resection is at active. In deed, if one is able to resect enough of the mass one might be able to man age the hy dro cep.h a lus di rectly recty and need for a ven tic u lar shunt. Fur ther more, as a gen er al prin ci ple of tu mor treat ment, debulking of the tu mor is thought to be posi tive. His tori cal ly, tu mors that were felt to be in trin sic in the op tic chiasm and hy pothal amus were con sid ered in op er a ble and the treat ment many years ago was to give ra di a tion ther apy as the only avail able op tion. With the ad vent of ad di tional tools such as the cav itron ul tra sonic sur gi cal as pir a tor (CUSA), re sec tion of op tic chiasmatic/hy pothal amic tu mors was at tempted and was com pleted suc cess ful ly.

In many re sec tions this type of sur gery is sim i lar to re sec tions of low grade astrocy tomas in the thalamus and spinal cord. As these pro ce dures be came more pop ular it be came increas ingly recog nized that there was a sig nif i cant mor bid ity asso ci ated with re sec ting tu mors from the chiasmatic/hy pothal amic re gion, par ti cul arly if an aggres sive sub to tal re sec tion was at tempted. Our ex pe rience, which is not dif fer ent from oth ers, is that these re sec tions may be as soci ated with a num ber of com pli ca tions such as in creased vi sual loss, vas cu lar in jury to per for a tion of ves sels with sub se quent stroke, hy pothal amic dam age and pi tu itary dys func tion. One of the major prob lems has been acute diffi culty with man age ment of wa ter and elec tro lyte bal ance in the first few days af ter the sur gi cal pro ce dure. In a re view of 18 pa tients with sur gi cal re sec tion for op tic chiasmatic/hy pothal amic tu mors at B.C. Children’s Hos pi tal, im me di ate post-operative di a betes insipidus was noted in 8 cases and 7 pa tients de vel op ed a hypona tre mia 1 to 5 days post-op era tively. Five of these chil dren with hy po na tre mia had sei su res re lated to their elec tro lyte bal ance: 1 went on to die and 2 were left with per ma nent mor bid ity fol low ing the hy po na tre mic sei zu res. Eight pa tients of the 18 went on to have per ma nent sei su res. Asymp tom atic in far c tion of fron tal lobe and deeper struc tures were seen com monly. The sig nif i cant mor bid ity asso ci ated with re sec ting of the su pe rior com ponent of the tu mor is clear at time of sur gery. We would con sider sur gi cal re sec tion of the su pe rior com ponent of the tu mor extend ing into the third ven tri cle and block ing the fo ramen Mon ro as a method of de com press ing the ven tri ce nu lar sys tem and avoid ing a shunt, with no at tempt be ing made to rad i cal re sec tion of the tu mor. In the first in stance, the sur gi cal pro ce dure is done by a stan dard pterional ap proach or subfrontal ap proach. In the lat ter sit u a tion an interhem ispheric ap proach through the cor pus cal lo sum is used to achieve the re sec tion of the su pe rior part of the tu mor. When a sig nif i cant amount of tu mor has been re sec ted (more than 25%), close ob serv a tion is re com med ed with out any ad juvant ther a py, with the ex cep tion of in fants. This excep tion re lates to the relatively rapid growth of op tic chiasmatic/hy pothal amic tu mors in the young pop u la tion, par tic u larly in in fan ts. We have ob served at least one in fant, in whom the tu mor grew back to al most the orig i nal size within 6 months of a sub to tal re sec tion (> 90% re sec tion), de spite the typ i cal his to logy of a pilocytic astrocytoma. Thus, in these youn ger chil dren, che mo ther a py, as dis cussed be low, should be con sid ered as an adju vant treat ment follow ing re sec tion, even if a large sur gi cal re sec tion has been achieved.

The gen er al trend for man age ment of op tic chiasm atic/hy pothal amic tu mors has been away from rad i cal re sec tion to war d the use of che mo ther a py as the first line of treat ment. Despite the fact that the tu mors are low grade astrocy tomas, che mo ther a py has been shown to be ef fec tive in this pop u la tion of pa tients. Ini tially, actino mycin D and vincristine were used in com bi na tion, but this has been su per seded by two more re cent com bi na tions. Current re com men da tions for pa tients with a de finite diag no sis of op tic chiasm atic/hy pothal amic as trocytoma would be a trial of one of the two che mo ther a peutic reg i mens. If the tu mor pro gresses on che mo ther a py, sur gi cal re sec tion might then be re com med ed. Ra dio ther a py is usu ally re com med ed only after at tempt ed che mo ther a py and sur gi cal re sec tion and par ti cu larly for older chil dren.

Ra dio ther a py used to be the main stay of treat ment for the op tic chiasm atic/hy pothal amic tu mors. How ever, it has be come re cognized that there are sig nif i cant neg a tive con se quences to ra dio ther a py, par ti cu larly in the
younger children in whom the optic chiasmatic/hypothalamic gliomas tend to occur. These complications have included intellectual changes related to radiation of the frontal and temporal lobes, endocrine dysfunction, visual loss, moyamoya syndrome and the development of secondary malignancies. It is recognized that radion therapy may have a positive effect on the tumor, but be cause of the noted negative consequences radiotherapy is generally withheld until other options have been exhausted. With newer forms of radiotherapy, including conformal radiotherapy and stereotactic radiotherapy, the role for radiotherapy may need to be revisited.

Conclusions
Optic pathway gliomas represent a heterogeneous group of tumors with different natural histories, depending on whether the tumor arises in the optic nerve, is localized to the optic chiasm or involved the optic chiasm and hypothalamus. In the past, these tumors have often been lumped together, thus creating significant confusion with respect to the expected natural history. If one excludes the intraorbital optic nerve tumors, it is still important to recognize that there is a difference, biologically, between the chiasmatic gliomas and the optic chiasmatic/hypothalamic gliomas. The chiasmatic tumors tend to be asso ciated with NF1, be have very much like a hamartoma, and can be managed by observation. Optic chiasmatic/hypothalamic tumors on the other hand, tend not to be associated with NF1, usually grow progressively, and require active therapeutic intervention. The first line of treatment is generally chemotherapy, with surgery reserved for progressive disease on the mother ap or to decompress the optic apparatus and/or ventricular system in certain cases. Radiation is probably best avoided, because of the significant associated morbidity. Radiotherapy is usually recommended as a last option, but newer therapeutic strategies may bring radiotherapy into greater favor.

REFERENCES


