#### NATIONAL COORDINATING COUNCIL FOR CANCER SURVEILLANCE BRAIN TUMOR WORKING GROUP 2004 REPORT

#### National Coordinating Council for Cancer Surveillance - Brain Tumor Working Group

In 2001, the National Coordinating Council for Cancer Surveillance (NCCCS) directed the Brain Tumor Working Group (BTWG) to provide implementation guidelines in order to proceed with Recommendations 3 and 4 contained in the BTWG's summary report, *Surveillance of Primary Intracranial and Central Nervous System Tumors: Recommendations from the Brain Tumor Working Group* (September, 1998; www. naaccr.org). Recommendations 3 and 4 state:

- 3. We recommend collection of data for primary intracranial and extracranial central nervous system (CNS) tumors by all registries, hospital-based, and population-based. This effort will necessitate a change in Commission on Cancer (COC) requirements and will increase costs to the hospital-based programs. Federal funding should be allocated to supplement the additional transition and ongoing data collection costs that will be incurred by central registries. Before additional data collection is implemented, a pilot study should be conducted in multiple states to assess the procedures and quality control functions needed, as well as the costs of collecting data on these tumors.
- 4. We recommend that the appropriate government and professional organizations be involved in carrying out the development and implementation of special training programs and curricula for central registry, hospital registry, and laboratory personnel, as well as the development of computerized edit-checking procedures. Training for reporting and tabulating primary intracranial and CNS tumors should be offered on a regular basis.

The BTWG is comprised of individuals who represent all cancer registry standard setting organizations as well as organizations and individuals interested in brain tumor surveillance. The organizations represented include the Central Brain Tumor Registry of the United States (CBTRUS), Centers for Disease Control's (CDC) National Program of Cancer Registries (NPCR), National Cancer Institute's (NCI) Surveillance, Epidemiology, and End Results (SEER) Program, American College of Surgeon's Commission on Cancer (COC) and the North American Association of Central Cancer Registries (NAACCR) Other members represent hospital and state cancer registries, software vendors, epidemiologists, and neuropathologists.

Many of the BTWG members are also members of various NAACCR committees and subcommittees. Members of the BTWG are interested in promoting the collection of non-malignant brain/CNS tumors and encouraging the development of the infrastructure to accomplish it. The BTWG provides a forum for synergy and communication both for its own charge and for the broader charges of Recommendations 3 & 4 (Appendix A - chart of relationships). Fulfillment of these recommendations is dependent on cooperation between multiple interdependent, independent organizations, and individuals.

## I. SUMMARY OF ACCOMPLISHMENTS

#### **SEER Special Studies**

SEER conducted special studies in New Mexico (1998, 2002), University of Southern California - Los Angeles (1998, 2002), Iowa (2000), and Utah (2002) to evaluate the collection of non-malignant brain/CNS tumors. SEER funded these special studies to address issues of measuring the extra

workload and to determine whether other case finding mechanisms would have to be used. Their findings identified in one study, over one-third of the non-malignant diagnoses were not histologically confirmed; there was only a radiographic diagnosis of the non-malignant tumor. Some studies concluded that there is approximately the same number of nonmalignant tumors as malignant ones. If the study is limited to histologically confirmed tumors, the numbers of non-malignant and malignant are similar, but if there is an effort to collect the non-histologically confirmed tumors, there are more non-malignant tumors than malignant. Non-malignant tumors occur more frequently than malignant tumors for acoustic nerve tumors, pituitary gland tumors, meninges, and in women, and persons over the age of 65.

#### **Passage of Legislation**

The North American Brain Tumor Coalition along with members of the brain tumor advocacy community mounted a grassroots campaign to establish federal legislation mandating the collection of non-malignant brain and central nervous system (CNS) tumors. On October 10, 2002, the Senate and House unanimously passed The Benign Brain Tumor Cancer Registries Amendment Act. After being signed by President Bush, this Act became Public Law 107-260 and requires the CDC National Program of Cancer Registries to collect all primary non-malignant brain and intracranial and central nervous system tumors (see Appendix B - Law).

#### **Reporting of Non-malignant Brain/CNS tumors**

The COC, SEER Program, and NPCR require the collection of non-malignant brain tumors with behavior codes of /0 or /1 for the following ICD-O-3 topography codes: C70.0-C72.9, C75.1-C75.3 for cases diagnosed in 2004 and forward.

#### **Canadian Council of Cancer Registries**

Collection of non-malignant intracranial and CNS tumors is not mandatory in Canada. Some provinces/territories currently collect C70.0 - C72.9 but not C75.1 - C75.3. Implementation of non-malignant intracranial and CNS tumor reporting would affect case ascertainment practices, case completeness, policies and procedures (nationally and provincially), workload, training, edits, software upgrades, etc. Canada is not adopting these guidelines at this time. The Canadian Council of Cancer Registries will discuss this further. (Source:

NAACCR 2004 Implementation Guidelines: Collaborative Staging and Benign/Borderline Intracranial and CNS Tumors, www.naaccr.org).

#### **Development and Approval of Non-malignant Brain Tumor Reporting Rules**

The NAACCR Registry Operations Committee (ROC) appointed a subcommittee for development of non-malignant brain tumor reporting rules in August 2002. This group included an expert neuropathologist as well as representatives from all the standard setting organizations, CBTRUS, and cancer registrars working in hospital and central cancer registries. Together they developed the document, *Accessioning Primary Intracranial and Central Nervous System Tumors: General Reporting Rules,* which were approved by NAACCR Uniform Data Standards (UDS) Committee in July 2003. These rules include coding laterality and assigning sequence numbers for non-malignant brain/CNS tumors along with new rules for determining multiple primaries for non-malignant brain/CNS tumors. Non-malignant brain/CNS tumor rules were implemented with 2004 diagnoses.

#### **Multiple Primaries**

Timing rule: The current timing rule for malignant multiple primary tumors is two months. For non-malignant brain/CNS tumors, a decision was made to eliminate the timing rule. The SEER Histology Committee is considering changes to the current two-month timing rule for multiple malignant tumors (see discussion later).

**Non-malignant tumors:** The development of the new multiple primary rules for nonmalignant brain/CNS tumors are based on the premises that the natural biology of nonmalignant tumors is that of expansive, localized growth, and local recurrence(s) is (are) common, and metastasis is uncommon or unusual. Non-malignant tumors of the same histology, same site, and same side will recur in the same location. If they recur, even after 20 years, they are still the same tumor. The summary of the rules is 1) multiple nonmalignant tumors of the same histology identified in the same location or side of the brain/CNS should be considered the same primary; 2) multiple non-malignant tumors of the same histology identified in different locations or sides of the brain/CNS should be considered separate primaries; and 3) multiple non-malignant CNS tumors with different histologies should be considered separate primaries.

**Definitions of same histology and different histology for non-malignant tumors:** When the ROC Brain Tumor subcommittee reviewed the current multiple primary rules to determine if they were applicable to non-malignant CNS tumors, they found that the current rules for "same/different histology" were not always applicable to non-malignant brain/CNS tumors. The current rule states: Differences in histologic type refer to differences in the first three digits of the morphology code. Some non-malignant brain/CNS tumor histologies do not follow this standard three-digit histology difference rule. Tumors may have histologies with differences in the first three digits (and sometimes even the first two digits) that actually represent a progression, differentiation or subtype of a single histological category. In other cases, tumors have the same first three digits of their histology codes, but are in fact very different biologically and should be counted as separate primary tumors. It became apparent that our biological understanding of brain/CNS tumors has changed over time and that the code groupings listed in the ICD-O are no longer considered to be biologically related. Using the current three-digit rule will result in some tumors being missed and others being over counted. For these reasons the Subcommittee recommended changes to the rules to more accurately represent the number of new brain/CNS tumors diagnosed. A table was developed that describes which non-malignant histology codes do not follow the standard three-digit histology difference rule and should be grouped together. (Appendix C)

**Malignant brain/CNS tumors:** The current multiple primary rules are retained for now. The ROC has proposed new rules to the SEER Histology Committee for implementation 1/1/2006 (see SEER Histology Committee).

# Laterality

The ROC proposed that laterality be collected on many brain/CNS sites for both malignant and non-malignant tumors. Laterality is necessary to determine multiple primaries for nonmalignant brain/CNS tumors where laterality is appropriate. The research community has indicated that the location and laterality for primary brain/CNS tumors is of significant interest in determining causation and assessing the impact on quality of life. (*Inskip PD*, *Neuroepidemiology 2003; 22;130-138*). Non-treatment-related factors such as location of tumor by hemisphere can be a predictive factor for cognitive outcome. (*Brown PD, Buckner, JC, Uhm JH, and Shaw EG (2003) The neurocognitive effects of radiation in adult low-grade* glioma patients. Neuro-Oncology 5, 161-167, 2003).

## **Changes to Data Collection Rules and Coding Structures**

Over the past several years, the NAACCR Uniform Data Standards Committee (UDSC) has been the vehicle through which the different organizations and committees have discussed new rules, changes to old rules, and changes to data items in order to lay the foundation for the collection of non-malignant brain/CNS tumors.

ICD-O-3 included pilocytic astrocytoma with a '1' behavior code and therefore, it would not be reportable beginning with 2001 diagnosis. This was discussed at the UDSC and the standard setters agreed to change their reporting requirements to include pilocytic astrocytomas. The behavior code for these cases is changed to '3'.

The BTWG discussed the impact of including non-malignant brain/CNS tumors. A summary of this discussion was presented to the UDSC in October 2002.

The SEER Summary Stage 1977 and 2000 did not have a code for non-malignant brain/CNS. An '8' (not applicable) was added to both SEER Summary Stage 1977 and SEER Summary Stage 2000 to handle the non-malignant brain/CNS cases. These changes were approved by the NAACCR Uniform Data Standards Committee.

Sequence number central and sequence number hospital were revised to handle nonmalignant cancers. The sequence number changes were implemented for cases diagnosed in 2003 forward. In July 2003, the UDSC accepted the proposed rules for non-malignant multiple brain/CNS tumors and changes in the sites for which laterality is coded for selected brain/CNS sites for both malignant and non-malignant tumors. The ROC document presented to UDSC also included guidelines for coding.

#### **Development of Edits**

Susan Capron and the NAACCR Edits Work Group have reviewed and amended NAACCR GenEDITS edits metafiles (versions 10 through 10C) during the last two years to accommodate the 2004 reporting requirements for intracranial non-malignancies and the parallel changes in data standards established for intracranial malignancies. For hospital and central registries choosing to collect intracranial non-malignancies in the past, and often for their software vendors, that choice meant having to modify and over-ride several standard edits designed to handle only malignancies. They will no longer have to write their own modifications and manually set those over-rides for diagnoses made beginning in 2004.

Over thirty standard edits\* were affected. Because many COC edits have accommodated the collection of non-malignancies for some time, the 2004 collection requirement by all standard-setters caused fewer overall edit changes than the data standards recently developed for intracranial tumors. A few new edits were created, many existing edits needed modification, and a few edits were found to no longer be needed as standard-setters came into agreement on certain practices. Some edits were altered multiple times as potential problems were recognized and data specifications became clearer.

Edits involving hospital and central sequence numbers were adjusted to accommodate the new separate numeric code ranges for sequencing non-malignancies in the 2003 data standards (version 10-10A metafiles). All other edit changes were made for 2004 implementation (version 10-10C metafiles). Diagnosis year-dependent considerations had to be incorporated into many edits.

With the exception of certain non-reportable malignancies and histologies which changed behavior (invasive to borderline, or borderline to invasive) between ICD-O-2 and ICD-O-3 coding, non-malignant behavior codes are limited to use with only the appropriate sequence number ranges specified in the data standards. Also with these certain exceptions, non-malignant behavior codes are permitted with only the appropriate intracranial primary site codes for diagnoses made beginning in 2004.

The altered standards for coding tumor laterality for some intracranial primary sites resulted in many adjustments to interfield edits involving the laterality codes.

Specific histologic type codes which formerly required edit over-ride flags to be set when assigned appropriately to intracranial non-malignancies will now pass morphology edits without review. Checks for unusual combinations of primary site and histology warranting review used to only include histologic type codes that are normally malignant when recorded

in most cancer data systems; purely non-malignant histologic type codes were added for the intracranial primary sites.

Validity checks on SEER general summary stage fields were modified to handle the code newly designated for non-malignancies.

The edits committee suggests that all Collaborative Staging fields be validated including the primary tumor extension code for intracranial non-malignancies and "CS Site Specific Factor 1" which contains the WHO grade for intracranial tumors.

Several edits only truly applicable to malignancies were made to disregard (skip) records in which sequence numbers, behavior or histologic type indicates a non-malignancy.

In some instances, updating a SEER edit for 2004 requirements made that edit exactly like an older COC edit that already accommodated intracranial non-malignancies. One of the redundant edits was deleted in such cases and sets of edits used by different groups and standard-setters were adjusted accordingly.

# NAACCR Brain/CNS Tumor Implementation Group

On November 7, 2003, NAACCR announced that the NAACCR 2004 Implementation Guidelines were available on the NAACCR web site. These guidelines include recommendations for the implementation of the Collaborative Staging (CS) System and the Reporting of Benign and Borderline Intracranial and CNS Tumors. Information in the Guidelines includes standard setting organization reporting requirements, Canadian Council of Cancer Registries position on non-malignant brain/CNS tumor collection, central registries implementation needs, reporting facilities implementation needs, software vendors' implementation needs, EDITS, training/education and appendices including Benign Brain Tumor Cancer Registries Amendment Act, Public Law 107-260; general reporting rules; ICD-9 and ICD-10 casefinding list; and, SEER ICD-O-3 Brain and CNS Site/Histology Type Validation list. These guidelines were updated on November 14, 2003 and errata added on February 2004 and are available on www.naaccr.org.

#### **Collaborative Staging Task Force**

The collection of non-malignant brain/CNS tumors impacted the Collaborative Staging in several ways. The American Joint Committee on Cancer (AJCC) 6<sup>th</sup> edition does not have a TNM or AJCC stage for brain/CNS tumors. A code '05' was added to the CS Extension field to accommodate non-malignant brain/CNS tumors. For brain/CNS tumors, WHO grade is important and the WHO grade code was added to CS Site Specific Factor 1. The NAACCR ROC committee proposed that syndromes be added to a site-specific factor in CS. It was decided that this did not fit into the timeframe of data collected under CS and therefore, was referred to UDS Committee.

#### **Educational Materials and Training Opportunities**

Training materials for Data Collection of Primary Central Nervous System Tumors were developed through the collaboration of the National Program for Cancer Registries (NPCR), SEER, NAACCR, and COC. In addition to NPCR, these materials have been approved by the American College of Surgeon's Commission on Cancer; NCI's SEER program, and NAACCR. All cancer registry standard-setting organizations have agreed to use these training materials to promote consistency in training.

In September 2003, NPCR offered a Cancer Registry Instructors Training Course in Atlanta, Georgia. The course was co-sponsored by NCI SEER, AJCC, and NAACCR. The course included presentations on Teaching Techniques for Effective Adult Learning, Data Collection of Primary CNS Tumors, and Understanding the Collaborative Staging System. Participants represented the staff in NPCR and SEER states responsible for providing training to cancer reporters in their states. Representatives from all but two NPCR states and all SEER states attended. Feedback from participants in that program and from state trainers was incorporated into these materials and they were distributed on a CD to these agencies and to all NPCR and SEER states cancer registries. The materials were posted on the NPCR website (http://www.cdc.gov/cancer/npcr/training/index.htm) in May 2004 and can be downloaded for use in local training workshops. These training materials have been used by state central cancer registry staff to train data collectors in their respective state/regional registries.

Data Collection of Primary CNS Tumors materials include a PowerPoint presentation with extensive speakers notes, exercises and answers suitable for printing, and other supporting documentation including:

- A. Benign Brain Tumor Cancer Registries Amendment Act
- B. Accessioning Primary Intracranial and Central Nervous System Tumors General Reporting Rules
- C. Additions to Cancer Registry ICD-9-CM and ICD-10 Casefinding List
- D. ICD-O-3 Primary Brain and CNS Site/Histology Listing
- E. Surveillance of Primary Intracranial and Central Nervous System Tumors: Recommendations from the Brain Tumor Working Group: National Coordinating Council for Cancer Surveillance
- F. Glossary of Terms: Intracranial and Central Nervous System (CNS) Tumors
- G. Presentation References
- H. Other References

SEER has prepared the following resources: 1) "Benign Brain Tumor Reporting", 2) The Brain Book, and 3) other presentations/training materials. All material is available to any interested parties.

1). "Benign Brain Tumor Reporting" is a web-based module (http://training.seer.cancer.gov) and provides the rationale for reporting non-malignant brain/CNS tumors, the background on the legislation, discussion of reportable cases and casefinding sources, and a discussion of the anticipated impact. The module continues with a discussion of the signs and symptoms, anatomy of the central nervous system, and other information data collectors need to abstract all types of CNS lesions. The module concludes with hands-on exercises including answers and rationales.

2). In a format similar to the Abstracting and Coding Guide for the Hematopoietic Diseases, SEER staff has developed a fascicle, The Brain Book, describing various aspects of abstracting central nervous system histologies. The information includes histologic terms, synonyms, casefinding codes, WHO grade, sites affected, symptoms, syndromes, likelihood to have radiation and/or chemotherapy and clinical/research notes. Coding guidelines are documented in the introduction. This draft is available in PDF or hard copy by contacting <u>april.fritz@nih.gov</u>. The Brain Book (currently in Draft 2 status, is not anticipated for publication until FY 2005).

3). Presentations and training materials prepared for CNS tumors and CNS tumor coding guidelines are available on the SEER web site, www.training.seer.cancer.gov. The presentations cover the anatomy, physiology and function of the CNS and the rules and guidelines for coding both malignant and newly reportable non-malignant tumors. Training materials include exercises adapted from the CDC/Atlanta training materials and independently developed exercises on determining multiple primaries within the CNS and other topics.

# II. PENDING ISSUES

#### **Registry Operations Committee Recommendations for Changes to the Multiple Primary Rules**

The NAACCR ROC Subcommittee proposed new rules for determining multiple primaries for malignant brain/CNS tumors. As they reviewed the current multiple primary rules to determine if they were applicable to benign tumors, they found that in addition to not being applicable to the non-malignant CNS tumors, the current rules for "same/different histology" were also not always applicable to malignant brain/CNS tumors. The Subcommittee recommended changes to the rules for malignant CNS tumors to more accurately represent the number of new brain/CNS tumors diagnosed. A table was developed that describes which malignant histology codes do not follow the standard three-digit histology difference rule and should be grouped together. These recommendations were not approved by UDS. UDS recommended that these rules be forwarded to the SEER Histology Coding Committee.

#### SEER Histology Coding Committee

The SEER Committee has been charged with review and revision of histology coding rules as well as SEER multiple primary coding rules for all sites. The SEER Histology Committee is composed of members from various organizations: CDC, NCI, COC, NAACCR, state registries, and pathologists. Subcommittees were formed for different disease sites.

The SEER committee began their review of the malignant rules proposed by the NAACCR ROC subcommittee on December 8, 2003. Membership was expanded to include Dr. Roger McLendon, neuropathologist, and the ROC subcommittee. By including the recommended malignant brain/CNS tumor rules in this process, it will allow rules changes to be implemented along with all other multiple primary rules for malignant tumors in 2006. Currently, the rules have been approved and are being revised into the committee's template being utilized for all sites. The committee plans to submit any rule changes to NAACCR UDS for approval and implementation for cases diagnosed 1/1/2006 and later.

#### **Recommendations from the Consensus Conference II**

A Consensus Conference on Brain Tumor Definition (Consensus Conference I) was facilitated by the Central Brain Tumor Registry of the United States (CBTRUS) and held on November 10, 2000 in Chicago, Illinois with the objective of reaching a multidisciplinary agreement on a standard brain tumor definition for the collection and comparability of data in the United States (McCarthy et al., Neuro-Oncology 4, 134-145, 2002). These objectives were guided by the first two recommendations of the Brain Tumor Working Group, a multidisciplinary team appointed by the National Coordinating Council for Cancer Surveillance in 1998 to investigate the collection of primary intracranial and central nervous system (CNS) tumors. Consensus was reached on the collection of all primary brain/CNS tumor histologies, including those with behavior coded benign and uncertain, as well as malignant, found and reported in the brain or CNS, International Classification of Disease for Oncology (ICD-O) site codes (C70.0-C72.9 & C75.1-C75.3). In addition, a comprehensive listing of those histologies occurring in the brain and CNS based on the CBTRUS grouping scheme was formulated to provide a template for reporting.

The Consensus Conference on Cancer Registration of Brain and Central Nervous System Tumors (Consensus Conference II) was held in November 2003 in Keystone, Colorado. Several issues identified during Consensus Conference I (McCarthy et al. 2002) or during the process of developing training materials for non-malignant brain/CNS tumor collection were tabled for future discussion. The CBTRUS facilitated a discussion between epidemiologists, neurosurgeons, and neuropathologists regarding these tabled issues during Consensus Conference II. Multidisciplinary consensus was reached on the following recommendations:

- (1) Change to cysts and tumor-like lesions that currently have an ICD-O-3 code, specifically that Rathke Pouch Tumours be given a separate ICD-O code rather than be combined with craniopharyngioma (9350/1),
- (2) Collection of additional cysts and tumor-like lesions that are located in brain and CNS sites but currently lack ICD-O codes; nine new codes are recommended,

- (3) New ICD-O topography site for skull base tumors for the brain and CNS (recommended new site code C70.2) to accurately capture tumours of the nervous system with increased topographic specificity, and
- (4) Collection genetic syndromes associated with CNS tumors.

Recommendations 1–3 of Consensus Conference II were sent to the International Association for Research on Cancer (IARC) and will be considered by the IARC Editorial Committee for revisions to the next edition of ICD-O. The IACR Editorial Committee has also received recommendations from other groups and all will be reviewed when the committee reassembles to discuss the next ICD-0 revision. CBTRUS will track any actions taken by the IARC Editorial Committee to the recommendations from the Consensus Conference. Recommendation 4 of Consensus Conference II has been sent to the ROC Subcommittee and has been reviewed by UDSC. UDSC has recommended that the concept of collecting genetic syndromes go to the NCCCS via the BTWG for further consideration. (Appendix D) CBTRUS will monitor and facilitate, when appropriate, action on these recommendations.

- Appendix A: BTWG activity flow chart
- Appendix B: Benign Brain Tumor Cancer Registries Amendment Act
- Appendix C: Table: Histologic groupings to determine same histology for non-malignant brain tumors
- Appendix D: August 29, 2003: Coding Genetic Syndromes: NAACCR Uniform Data Standards Committee response to Registry Operations Committee

# Appendix A Activities Leading to Implementation of Reporting Benign Brain Tumors 1997 to 2002



# Appendix A, (continued) Activities Leading to Implementation of Reporting Benign Brain Tumors 2003 to 2004



# **APPENDIX B**

## **SECTION 1. SHORT TITLE.**

This Act may be cited as the `Benign Brain Tumor Cancer Registries Amendment Act'. SEC. 2. NATIONAL PROGRAM OF CANCER REGISTRIES; BENIGN BRAIN-RELATED

TUMORS AS ADDITIONAL CATEGORY OF DATA COLLECTED. (a) IN GENERAL- Section 399B of the Public Health Service Act (42 U.S.C. 280e), as redesignated by section 502(2) (A) of Public Law 106-310 (114 Stat. 1115), is amended in subsection (a)--

(1) by redesignating paragraphs (1) through (5) as subparagraphs (A) through (E), respectively, and indenting appropriately;

(2) by striking `(a) IN GENERAL- The Secretary' and inserting the following: `(a) IN GENERAL-

`(1) STATEWIDE CANCER REGISTRIES- The Secretary';

(3) in the matter preceding subparagraph (A) (as so redesignated), by striking `population-based' and all that follows through `data' and inserting the following: `population-based, statewide registries to collect, for each condition specified in paragraph (2)(A), data'; and

(4) by adding at the end the following:

(2) CANCER; BENIGN BRAIN-RELATED TUMORS-

`(A) IN GENERAL- For purposes of paragraph (1), the conditions referred to in this paragraph are the following:

`(i) Each form of in-situ and invasive cancer (with the exception of basal cell and squamous cell carcinoma of the skin), including malignant brain-related tumors.

`(ii) Benign brain-related tumors.

`(B) BRAIN-RELATED TUMOR- For purposes of subparagraph (A):

`(i) The term `brain-related tumor' means a listed primary tumor (whether malignant or benign) occurring in any of the following sites:

`(I) The brain, meninges, spinal cord, cauda equina, a cranial nerve or nerves, or any other part of the central nervous system.

`(II) The pituitary gland, pineal gland, or craniopharyngeal duct.

`(ii) The term `listed', with respect to a primary tumor, means a primary tumor that is listed in the International Classification of Diseases for Oncology (commonly referred to as the ICD-O). `(iii) The term `International Classification of Diseases for Oncology' means a classification system that includes topography (site) information and histology (cell type information) developed by the World Health Organization, in collaboration with international centers, to promote international comparability in the collection, classification, processing, and presentation of cancer statistics. The ICD-O system is a supplement to the International Statistical Classification of Diseases and Related Health Problems (commonly known as the ICD) and is the standard coding system used by cancer registries worldwide. Such term includes any modification made to such system for purposes of the United States. Such term further includes any published classification system that is internationally recognized as a successor to the classification system referred to in the first sentence of this clause. `(C) STATEWIDE CANCER REGISTRY- References in this section to cancer registries shall be considered to be references to registries described in this subsection.'.

(b) APPLICABILITY- The amendments made by subsection (a) apply to grants under section 399B of the Public Health Service Act for fiscal year 2002 and subsequent fiscal years, except that, in the case of a State that received such a grant for fiscal year 2000, the Secretary of Health and Human Services may delay the applicability of such amendments to the State for not more than 12 months if the Secretary determines that compliance with such amendments requires the enactment of a statute by the State or the issuance of State regulations.

# **APPENDIX C**

A.	Table 3.	Histologic groupings to determine same histology for non-malignant
		brain tumors

Choroid plexus neoplasms	9390/0, 9390/1
Ependymomas	9383, 9394, 9444,
Neuronal and neuronal-glial	9384, 9412, 9413, 9442, 9505/1,
neoplasms	9506
Neurofibromas	9540/0, 9540/1, 9541, 9550,
	9560/0
Neurinomatosis	9560/1
Neurothekeoma	9562
Neuroma	9570
Perineurioma, NOS	9571/0

# **APPENDIX D**

# August 29, 2003

# Coding Genetic Syndromes – NAACCR Uniform Data Standards Committee (UDS) response to Registry Operations Committee (ROC)

The UDSC has considered the recommendation from the ROC Benign Brain Tumor Subcommittee to collect information regarding specific genetic syndromes associated with brain and central nervous system tumors. While UDSC appreciates the clinical application of this information when describing brain/CNS tumor diagnoses the committee can not justify the addition of a new item to the NAACCR data transmission layout specifications for this specific purpose.

The UDSC recommends that the ROC BBTSC consult with NCCCS. This coordinating group should be asked to consider drafting a broader application of this concept. The development of a more widely applicable data construct should apply to cancers across a number of organ sites, not simply brain/CNS tumors, which have genetic syndromes or medical conditions associated with the diagnosis and/or outcomes of particular cancers.

UDSC requests that the following issues are considered if NCCCS elects to pursue such a project:

- 1) list of applicable disease sites and associated syndromes and/or conditions
- 2) availability of the data elements in patient medical records
- 3) reliability with which data elements can be recorded by registry professionals
- 4) particular coding guidelines necessary to record the data elements, including timing rules for data element abstracting and coding
- 5) estimates of the costs associated with time requirements and registry resources required to abstract these data elements
- 6) expected/potential application and use of the data elements
- 7) interest among standard setting organizations to sponsor and maintain these data elements
- 8) interest among standard setter organizations to require the abstracting and reporting of these data elements