

**COLLECTION AND CODING CLARIFICATIONS  
FOR CENTRAL NERVOUS SYSTEM (CNS) TUMORS**

Date of clarification: July 2004

Subject: **Collection and coding of blood vessel tumors occurring in CNS sites**

Question: Are non-malignant blood vessel tumors occurring in CNS sites reportable?

Answer: YES: The CNS site/histology listing includes blood vessel tumors under several categories including meninges C70.0 -C70.9, brain C71.0-C71.4 and C71.7-C71.9, (excluding ventricle), spinal cord C72.0, cauda equina C72.1, cranial nerves C72.2-C72.5, cerebellum C71.6 and other nervous system C72.8-C72.9.

These tumors include:

9120/0 Hemangioma, NOS  
9121/0 Cavernous hemangioma  
9150/0 Hemangiopericytoma, benign  
9150/1 Hemangiopericytoma, NOS  
9161/1 Hemangioblastoma

Question: Should blood vessel tumors occurring in CNS sites be coded to blood vessel or CNS?

Answer: They should be coded to the CNS site in which they occur.

This follows the same rationale in that if you have a lymphoma of the brain, it is coded to "brain," not lymph node.

*Example:*

Patient is diagnosed with a hemangioblastoma of the right temporal lobe of the brain. Site code: C71.2, histology code: 9161/1 Laterality: Right only

Date of clarification: March 2004

Subject: **Coding multiple meningiomas**

1. Cerebral meningiomas are coded to C70.0; Spinal meningiomas are coded to C70.1.
2. Cerebral meninges is listed as a lateral site, so you code **left** cerebral meninges and/or **right** cerebral meninges when possible.

*Example:* Patient has a meningioma over the right temporal lobe and a second meningioma over the left frontal area. Code the laterality of the first tumor to "right" cerebral meninges and the laterality of the second tumor to "left" cerebral meninges.

3. The morphology code for benign meningioma, NOS is M9530/0 (other benign codes are in the 953-series).
4. Use of the code for "multiple meningiomas," M9530/1

This has a behavior code of "1" and so does NOT apply to multiple or sequential benign meningiomas that have a behavior code of /0 OR **malignant** meningiomas that have a behavior code of /3.

Do not use this code for multiple meningiomas except in rare circumstances (about 1-2% of all meningiomas). Multiple meningiomas (also called **meningiomatosis**), is a rare condition strongly associated with neurofibromatosis type 2 and other genetic disorders, but occasionally occurs as sporadic or idiopathic. Occasionally multiple meningiomas may arise that are not associated with a genetic disorder. However, multiple malignant meningiomas are rare.

Subject: **Multiple primary rules for meningiomas**

- A. Laterality is coded on cerebral meninges. The benign CNS tumor rules indicate that if there are multiple tumors in "different sites" defined as cerebral (C70.0) versus spinal (C70.1) OR "different laterality" within cerebral meninges, they are **separate tumors**.

*Examples:*

1. Patient has a benign cerebral meningioma, NOS over the left parietal lobe. Site code: C70.0; Laterality: Left only, M9530/0. The patient is subsequently diagnosed with a benign cerebral meningioma, NOS on the right side of the brain. Site code: C70.0; Laterality: Right only; M9530/0. Because these benign tumors are located on different sides of the brain, they are considered **separate** primary tumors. Two abstracts should be completed.
2. Patient has a benign cerebral meningioma, NOS over the left parietal lobe. Site code: **C70.0**; Laterality: Left only, M9530/0. The patient is subsequently diagnosed with a benign spinal meningioma, NOS. Site code: **C70.1**; Laterality: Not a paired site; M9530/0. Because these tumors are located in different subsites, they are considered **separate** primary tumors. Two abstracts should be completed.

- B. Conversely, if they are in the "same site" (subsite and laterality), they are the **same tumor** regardless of the timing.

*Example:*

1. Patient has a benign cerebral meningioma, NOS over the left parietal lobe. Site code: **C70.0**; Laterality: Left only, M9530/0. The patient is subsequently diagnosed with a benign cerebral meningioma, NOS also on the left side of the brain. Site code: **C70.0**; Laterality: Left only; M9530/0. Because these tumors are in the same site (C70.0), same histology (M9530/0), and located on the same side of the brain, they are considered the **same** primary tumor regardless of the timing of the diagnosis. Only one abstract should be completed.

- C. If one meningioma is **benign** (/0 or /1) and one is **malignant** (/3), they are always considered separate tumors and two abstracts are completed.