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### Congenital tumors of the central nervous system.

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#### Abstract

Congenital tumors of the central nervous system (CNS) are often arbitrarily divided into "definitely congenital" (present or producing symptoms at birth), "probably congenital" (present or producing symptoms within the first week of life), and "possibly congenital" (present or producing symptoms within the first 6 months of life). They represent less than 2% of all childhood brain tumors. The clinical features of newborns include an enlarged head circumference, associated hydrocephalus, and asymmetric skull growth. At birth, a large head or a tense fontanel is the presenting sign in up to 85% of patients. Neurological symptoms as initial symptoms are comparatively rare. The prenatal diagnosis of congenital CNS tumors, while based on ultrasonography, has significantly benefited from the introduction of prenatal magnetic resonance imaging studies. Teratomas constitute about one third to one half of these tumors and are the most common neonatal brain tumor. They are often immature because of primitive neural elements and, rarely, a component of mixed malignant germ cell tumors. Other tumors include astrocytomas, choroid plexus papilloma, primitive neuroectodermal tumors, atypical teratoid/rhabdoid tumors, and medulloblastomas. Less common histologies include craniopharyngiomas and ependymomas. There is a strong predilection for supratentorial locations, different from tumors of infants and children. Differential diagnoses include spontaneous intracranial hemorrhage that can occur in the presence of coagulation factor deficiency or underlying vascular malformations, and congenital brain malformations, especially giant heterotopia. The prognosis for patients with congenital tumors is generally poor, usually because of the massive size of the tumor. However, tumors can be resected successfully if they are small and favorably located. The most favorable outcomes are achieved with choroid plexus tumors, where aggressive surgical treatment leads to disease-free survival.

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