Hypothalamic hamartomas (HH) are rare, tumor-like malformations that occur during fetal development and are present at birth. They are non-progressive lesions and do not expand, spread or metastasize to other locations. They grow in proportion to normal brain growth, and consequently their relative size to the rest of the brain is the same for the lifetime of the patient. There is tremendous diversity in the type and severity of symptoms from patient to patient. However, symptoms are apparent during childhood in the overwhelming majority of patients. Two clinical phenotypes of HH are recognized: 1) central precocious puberty and 2) epilepsy and related neurobehavioral symptoms. Hypothalamic hamartomas are relatively rare. Population-based research has shown that HH with epilepsy occurs in 1 of 200,000 children and adolescents. The prevalence of HH with only precocious puberty is unknown. At least for HH with epilepsy, males appear to have a slightly higher risk than females (approximately 1.3 to 1 ratio). HH occurs worldwide, without any obvious geographical concentration of cases. It is currently felt that all ethnic groups are at equal risk. There are no identified maternal risk factors or fetal exposures that increase the risk of HH. As malformations of the ventral (inferior) hypothalamus, hypothalamic hamartomas are rather distinct from other conditions. It is important to note that tumors that occur in the same region (such as craniopharyngiomas, astrocytomas, optic nerve gliomas, etc) usually do not cause gelastic seizures. These tumors can, however, be associated with endocrine dysfunction, which includes precocious puberty under rare circumstances.