

## Definition of Hypothalamic Hamartoma

Hypothalamic Hamartoma are very rare, congenital, benign brain tumour located close to or within the hypothalamus. Individuals who have HH may be asymptomatic but they are usually diagnosed because of its presentation; precocious puberty, Gelastic seizure or disruptive behavioural problems and cognitive deterioration.

- It has been estimated that incident of hypothalamic hamartoma ranges from 1/100,000 to 1/100,000,000.

## Classification of Hypothalamic Hamartoma (HH)

This classification of HH is a system that is based on size, hypothalamic displacement, origin and sit of attachment.

Type	Size	Attachment	Origin	Hypothalamic Displacement
Ia	Small-medium (diameter less than 1.5cm)	Pedunculated (parahypothalamic)	Tuber cinereum	No
Ib	Small-medium	Pedunculated (parahypothalamic)	Mammillary body	No
IIa	Medium-large (usually larger than 1.5cm)	Sissile (intrahypothalamic)	Tuber cinereum/mammillary body	Slight
IIb	Medium-large	Sessile (Intrahypothalamic)	Tuber cinereum/mammillary body	Marked

Valdueva et al.

\* Pedunculated HHs (parahypothalamic) have a narrow based attachment to the hypothalamus without any extension from the third ventricle and not typically associated with hypothalamic distortion. These have been more associated with precocious puberty.

\*sessile HHs (intrahypothalamic) have a broad based attachment, with or without extension from the third ventricle and may have some degree of hypothalamic distortion. Seizures have been found to be more common on this type. Other associated problems with sessile HHs are cognitive decline, developmental retardation, psychiatric problems; rage behaviour and mood liability.