

Primitive neuroectodermal tumor

In [1973](#), Hart and Earle coined the term “primitive neuroectodermal tumor” ¹⁾

Primitive [neuroectodermal tumors](#) (PNETs) are a group of highly [malignant tumors](#) composed of small round cells of neuroectodermal origin that affect soft tissue and bone. Primitive neuroectodermal tumors (PNETs) exhibit great diversity in their clinical manifestations and pathologic similarities with other small, round cell tumors. This has made classifying this family of tumors challenging and controversial. Batsakis et al (1996) divided the primitive neuroectodermal tumor (PNET) family of tumors into the following 3 groups based on the tissue of origin: ²⁾.

CNS primitive neuroectodermal tumors (PNETs) - Tumors derived from the central nervous system

[Neuroblastoma](#) - Tumors derived from the autonomic nervous system

[Peripheral primitive neuroectodermal tumors](#) (pPNETs) - Tumors derived from tissues outside the central and autonomic nervous system

The site of origin of primitive [neuroectodermal tumors](#) (PNETs) is quite varied and has significant influence on the prognosis.

Supratentorial primitive neuroectodermal tumor

[Supratentorial primitive neuroectodermal tumor](#).

Primary spinal peripheral primitive neuroectodermal tumor

see [Primary spinal peripheral primitive neuroectodermal tumor](#)

Brainstem primitive neuroectodermal tumor

Primitive neuroectodermal tumors of the central nervous system (CNS-PNET) arising in the brainstem are extremely rare, and knowledge about them is limited. The few existing case series report fatal outcomes.

Between September 1992 and November 2011, 6 eligible children with histologically proven brainstem CNS-PNET not otherwise specified and 2 children with brainstem ependyoblastomas (3, partial resection; 3, subtotal resection; 2, biopsy), median age 3.3 years (range 1.2-10.6 years), were treated according to consecutive multimodal HIT protocols for CNS-PNET/[medulloblastoma](#). Postoperative treatment was according to maintenance chemotherapy protocols (3, craniospinal irradiation [CSI] followed by maintenance chemotherapy), sandwich chemotherapy protocols (2, neoadjuvant chemotherapy, CSI, maintenance chemotherapy), or a therapy protocol for children younger than 4 years (3, postoperative chemotherapy followed by CSI).

The median duration of prediagnostic symptoms, predominantly cranial nerve deficits (n = 7),

pyramidal tract signs (n = 5), or ataxia (n = 5), was 5 weeks (range 1-13 weeks). The tumors were all located in the pons. Most involved more than half of the pontine axial diameter and were sharply margined. All patients had postoperative residual disease, including metastasis in 1 case. With 1 exception all tumors progressed early during treatment within 3.9 months (range 2.5-10.4 months), leading to a 1-year event-free survival rate (\pm standard error) of $13\% \pm 12\%$. After progression, patients succumbed early to their disease resulting in a 1-year overall survival rate of $25\% \pm 15\%$. The only surviving patient had a partially resected CNS-PNET, received a sandwich chemotherapy protocol, and is without disease progression 14 months after diagnosis.

CNS-PNET is a rare but important differential diagnosis in childhood brainstem tumors. So far, efficient therapies are lacking. The sampling of tumor material for improved biological understanding and identification of new therapeutic targets is important ³⁾.

1)

Hart MN, Earle KM. Primitive neuroectodermal tumors of the brain in children. Cancer. 1978;32:890-97.

2)

Batsakis JG, Mackay B, el-Naggar AK. Ewing's sarcoma and peripheral primitive neuroectodermal tumor: an interim report. Ann Otol Rhinol Laryngol. 1996 Oct. 105(10):838-43.

3)

Friedrich C, Warmuth-Metz M, von Bueren AO, Nowak J, Bison B, von Hoff K, Pietsch T, Kortmann RD, Rutkowski S. Primitive neuroectodermal tumors of the brainstem in children treated according to the HIT trials: clinical findings of a rare disease. J Neurosurg Pediatr. 2015 Mar;15(3):227-35. doi: 10.3171/2014.9.PEDS14213. Epub 2015 Jan 2. PubMed PMID: 25555122.

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