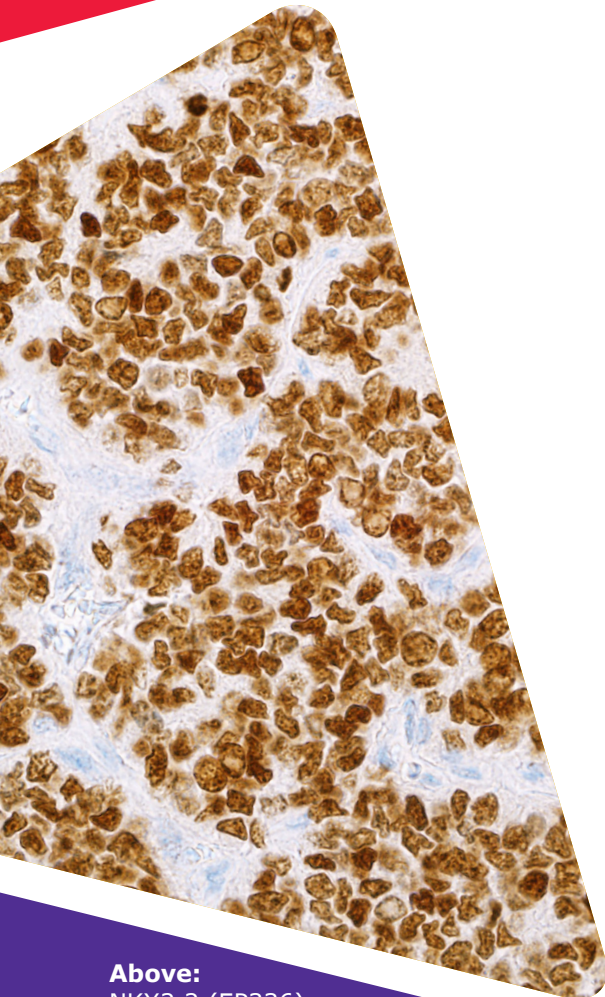


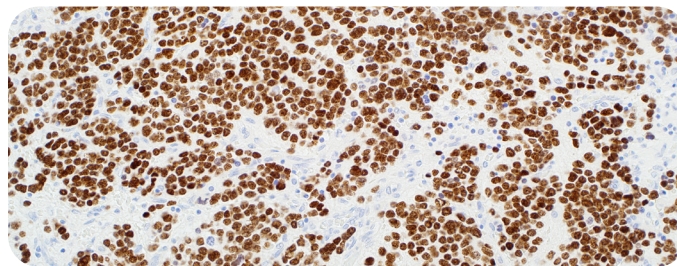
Cell Marque™ Tissue Diagnostics

Pediatric Pathology



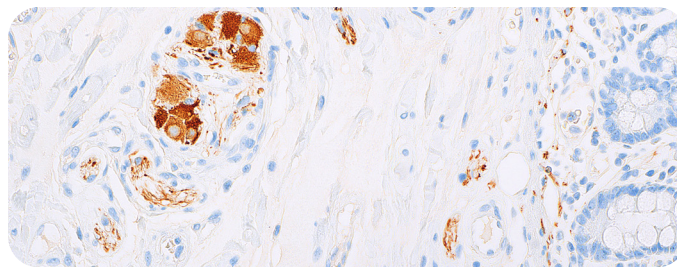
Above:
NKX2.2 (EP336)

View more at:
cellmarque.com/specialties



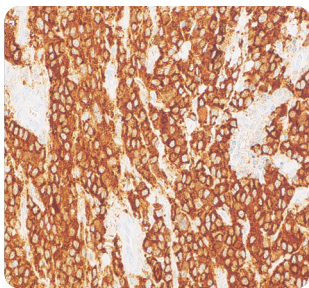
PHOX2B (EP312)

Paired-like homeobox 2B (PHOX2B) is a transcription factor located on chromosome 4p13 which is crucial to the formation of autonomic ganglia in the autonomic nervous system (ANS). PHOX2B gene is strictly expressed in neural crest derivatives committed to the noradrenergic phenotype. The PHOX2B gene encodes a paired-like homeo-domain transcription factor with an extra-axial expression pattern restricted to the ANS. Neuroblasts of peripheral neuroblastic tumors are derived from the sympathoadrenal lineage, a division of the ANS. PHOX2B has been observed in peripheral neuroblastic tumors, neuroblastomas, paragangliomas, ganglioneuroblastomas, ganglioneuromas and pheochromocytomas. PHOX2B has been reported to be negative in other small round blue cell tumors.



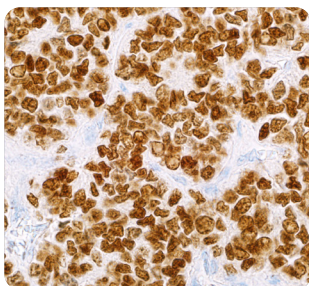
Peripherin (8G2)

Peripherin is a type III intermediate filament protein localized in the cytoplasm of ganglion cells and in a granular pattern in nerve fibers comprising the peripheral nervous system, which extends to many tissues throughout the body including the salivary gland, small intestine, prostate, stomach, and colon. The specific expression of peripherin in ganglia provides utility in the identification of immature ganglion cells in infant or newborn biopsies where there may be morphological ambiguity with other cell types such as endothelial cells, fibroblasts, or inflammatory cells. Likewise, peripherin labeling can be used for visualizing ganglion cell distribution where reduction or loss of observed labeling is correlative with the reduction or loss of ganglion cell presence. The use of anti-peripherin in tracking the reduction or loss of ganglion cells in the submucosal and myenteric layers of the colon wall can act as a valuable tool in identifying patients suspected of recto-sigmoid Hirschsprung disease and other forms of colonic aganglionosis.



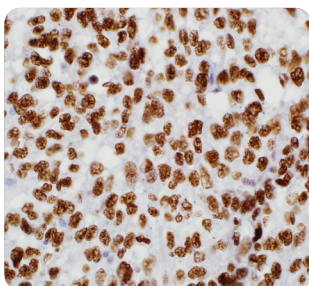
Tyrosine Hydroxylase (LNC1)

Tyrosine Hydroxylase (TYH) is the rate-limiting enzyme of catecholamine biosynthesis. TYH uses tetrahydrobiopterin and molecular oxygen to convert tyrosine to L-DOPA, a precursor to dopamine. Anti-TYH is sensitive and specific for peripheral neuroblastic tumors in the differential diagnosis from other tumors of childhood and shows high diagnostic utility. Anti-TYH has been shown to positively label all pheochromocytomas and sympathetic paragangliomas, and thus aids in differentiating them from their histologic mimics such as adrenocortical neoplasms.



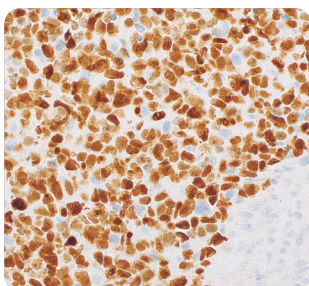
NKX2.2 (EP336)

NKX2.2 is a homeodomain transcription factor that plays a role in neuroendocrine and glial differentiation. NKX2.2 is upregulated in Ewing's sarcoma as a result of the oncogenic EWS-FLI1 fusion protein. As one of many small round blue cell tumors, Ewing's sarcoma can be difficult to diagnose due to the characteristic undifferentiated morphology. NKX2.2 has proven its utility as a sensitive marker for distinguishing Ewing's sarcoma from other round blue cell tumors when used in a panel.



TFE3 (MRQ-37)

Transcription factor E3 (TFE3) is a protein expressed in many cell types that are encoded by the TFE3 gene. This gene may be involved in chromosomal translocations that occur in some cancers. Xp11 translocation renal cell carcinomas (RCC) are a recently recognized subset of RCC, characterized by chromosome translocations involving the Xp11.2 break point and resulting in gene fusions involving the TFE3 transcription factor gene that maps to this locus. Alveolar soft part sarcoma (ASPS) is an uncommon soft tissue sarcoma of uncertain differentiation. The hallmark of ASPS is a chromosomal rearrangement at 17q25 and Xp11.2 engendering an ASPSCR1-TFE3 fusion gene responsible for an aberrant transcription factor presumably enabling pathogenesis.



PAX-7 (MRQ-69)

The paired-box (PAX) family of proteins are key transcriptional regulators involved in early critical development. The PAX-7 transcription factor has important functions in mammalian myogenesis and early neural development, with a particularly crucial role in specification and self-renewal of skeletal muscle tissue. A high frequency of strong PAX-7 nuclear expression has been identified predominantly in rhabdomyosarcomas, preferentially in the embryonal subtype, and Ewing's sarcoma, with reactivity being absent in related malignancies such as leiomyosarcoma, lymphoblastic lymphoma, neuroblastoma, carcinoid tumor, gastrointestinal stromal tumor, and small cell lung carcinoma. Immunohistochemical detection of PAX-7 protein can be used as a tool in distinguishing embryonal rhabdomyosarcoma and Ewing's sarcoma from histologic mimics.

The product featured belongs to the group *in vitro* diagnostic (IVD) medical devices. The product is classified as being IVD Class 1, exempt per US FDA regulation, and complies with the EU IVD Directive, bearing the CE logo on the label. The product featured is not available in all countries. Contact your local sales representative or distributor for details.

For ordering information, please contact your local sales representative or distributor.

For full references and product details please see the product insert.

These products herein are intended for laboratory use in the detection of their respective proteins in formalin-fixed, paraffin-embedded human tissue stained in qualitative immunohistochemistry (IHC) testing. These products are not a stand-alone diagnostic, and cannot be used for diagnosis, treatment, prevention, or mitigation of disease.

Direct: +1 916-746-8900
Fax: +1 916-746-8989
Email: international@milliporesigma.com
www.cellmarque.com

For sizes and availability please visit www.cellmarque.com.

Merck KGaA
Frankfurter Strasse 250
64293 Darmstadt, Germany

SigmaAldrich.com

CELL MARQUE
RabMab
Technology from Abcam

Copyright © 2022 Merck KGaA, Darmstadt, Germany and/or its affiliates. All Rights Reserved. Merck, the vibrant M, Sigma-Aldrich and Cell Marque are trademarks of Merck KGaA, Darmstadt, Germany or its affiliates. All other trademarks are the property of their respective owners. Detailed information on trademarks is available via publicly accessible resources.

MK_FL6657EN Ver. 2.0 34394 03/2022

