

PHOX2B (Paired Mesoderm Homeobox Protein 2B) (EP312)

Rabbit anti-human PHOX2B Monoclonal Antibody (Clone EP312)

REFERENCES AND PRESENTATIONS¹

- ready-to-use (manual or LabVision AutoStainer)
 MAD-000748QD–3
 MAD-000748QD–7
 MAD-000748QD–12
- Ready-to-use (MD-Stainer)² MAD-000748QD-3/V MAD-000748QD/V
- concentrated MAD-000748Q - 1:50 recommended dilution

COMPOSITION

Anti-human PHOX2B rabbit monoclonal antibody purified from culture supernatant, filtered, sterilized and prepared in 10mM PBS, pH 7.4, with 0.2% BSA and 0.09% sodium azide

INTENDED USE The Immunohistochemistry (IHC) on paraffin embedded tissues. Not tested on frozen tissues or Western-Blotting CLONE: EP312³

Ig ISOTYPE: Rabbit IgG

IMMUNOGEN: Synthetic peptide corresponding to the human PHOX2B.

SPECIES REACTIVITY: In vitro diagnostics in humans. Not tested in other species

DESCRIPTION AND APPLICATIONS:

The PHOX2B gene (also known as PMX2B or NBPHOX) is located in the chromosome region 4p13 and encodes a nuclear transcription factor that, during the embryonic development, predominantly expresses in the autonomic nervous system, being essential in the differentiation and survival of vegetative neurons and chromaffin cells. Mutations of the PHOX2B gene are responsible for the central hypoventilation syndrome, a syndrome with autosomal dominant transmission in which the hypoventilation is secondary to the

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Vitro S.A

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reduction or absence of ventilatory response to hypercapnia or progressive hypoxemia and, as a consequence, new born die during sleep. By definition, there must not be specific neuromuscular diseases of the central nervous system; metabolic, lung, heart diseases or other lesions that explain hypercapnia. This syndrome is associated with tumors of neural crest origin, including neuroblastoma, as well as Hirschsprung's disease because of the lack of development of the vegetative neurons of the myenteric plexuses.

In normal tissues, the nuclear positivity of PHOX2B is observed in the vegetative neurons during the whole embryonic development and continues in adults.

In the differential diagnosis with other small-blueround-cell tumors of childhood (such as Wilm's tumor, Ewing's sarcoma or rhabdomyosarcomas), the antibody has shown its specificity in all the stages of the differentiation of both the primary and metastatic neuroblastoma. Its nuclear staining allows an exact and easy interpretation, avoiding the staining artifacts and false positives of CD57 or Synaptophysin, especially in decalcified bone biopsies. PHOX2B is not only important in the diagnosis of neuroblastoma, but also in the staging and follow-up of the disease, since the staining is maintained despite of the fibrosis, abundant inflammation or diffuse calcification, sometimes observed in the post-treatment samples.

In a study including other neuronal or neuroendocrine tumors, all of them of neural crest origin, the antibody has shown staining in neuroblastomas, ganglioneuroblastomas and ganglioneuromas, as well as 40% of paragangliomas, whereas pheochromocytoma, Merkel-cell carcinomas. neuroendocrine carcinomas of the lung and the gastrointestinal tract, tumors with thyroid. parathyroid and adrenal cortical origin or melanomas have been negative.

IHC POSITIVE CONTROL: Tissue section from neuroblastoma or normal colon. VISUALIZATION: Nuclear

IHC RECOMMENDED PROCEDURE:

- $4\mu m$ thick section should be taken on charged slides; dry overnight at $60^{\circ}C$
- Deparaffinise, rehydrate and HIER (heat induced epitope retrieval) boil tissue in the Pt Module



¹ These references are for presentation in vials of Low Density Polyethylene (LDPE) dropper. In case the products are used in automated stainers, a special reference is assigned as follows:

 [/] L: Cylindrical screw-cap vials (QD-3 / L, QD-7 / L, QD-12 / L).
/ N: Polygonal screw-cap vials (QD-3 / N, QD-7 / N, QD-12 / N).

For different presentations (references / volumes) please contact the supplier.

² For Technical specifications for MD-Stainer, please contact your distributor.

³ CK17 clone EP98 is manufactured using Epitomics's RabMAb® technology under U.S. Patent Nos. 5,675,063 and 7,402,409



using Vitro S.A EDTA buffer pH8⁴ for 20 min at 95°C. Upon completion rinse with 3-5 changes of distilled or deionised water followed by cooling at RT for 20 min

- Endogenous peroxidase block Blocking for 10 minutes at room temperature using peroxidase solution (ref. MAD-021540Q-125)
- Primary antibody: incubate for 20 minutes [The antibody dilution (when concentrated) and protocol may vary depending on the specimen preparation and specific application. Optimal conditions should be determined by the individual laboratory]
- For detection use Master Polymer Plus Detection System (HRP) (DAB included; ref. MAD-000237QK)
- Counterstaining with haematoxylin and final mounting of the slide

STORAGE AND STABILITY: Stored at 2-8°C. Do not freeze. Once the packaging has been opened it can be stored until the expiration date of the reagent indicated on the label. If the reagent has been stored under other conditions to those indicated in this document, the user must first check its correct performance taking into account the product warranty is no longer valid.

WARNINGS AND PRECAUTIONS:

1. Avoid contact of reagents with eyes and mucous membranes. If reagents come into contact with sensitive areas, wash with copious amounts of water.

2. This product is harmful if swallowed.

3. Consult local or state authorities with regard to recommended method of disposal.

4. Avoid microbial contamination of reagents.

SAFETY RECOMMENDATIONS

This product is intended for laboratory professional use only. The product is NOT intended to be used as a drug or for domestic purposes. The current version of the Safety Data Sheet for this product can be downloaded by searching the reference number at <u>www.vitro.bio</u> or can be requested at <u>regulatory@vitro.bio</u>.

BIBLIOGRAPHY

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8. Lee JP, Hung YP, O'Dorisio TM, Howe JR, Hornick JL, Bellizzi AM. Examination of PHOX2B in adult neuroendocrine neoplasms reveals relatively frequent expression in phaeochromocytomas and paragangliomas. Histopathology. 2017 Oct;71(4):503-510.

LABEL AND BOX SYMBOLS

Explanation of the symbols of the product label and box:

\square	Expiration date
X	Temperature limit
***	Manufacturer
T	Sufficient content for <n> assays</n>



⁴ Ref: MAD-004072R/D – Vitro S.A



REF	Catalog number
LOT	Lot code
-T-1	Refer to the instructions of
Li	use
	Medical product for in
IVD	<i>vitro</i> diagnosis.
e-SDS	Material safety data sheet



