

Pendrin (G-19): sc-23779

BACKGROUND

Pendred syndrome (PDS), an autosomal recessive disorder, is the most common form of syndromic deafness characterized by congenital sensorineural hearing loss and goiter. The gene associated with PDS is mapped to chromosome 7 and encodes a putative transmembrane protein designated Pendrin. Several mutations in the gene have been identified and account for about 10% of hereditary deafness. Pendrin transcripts are expressed at significant levels in the thyroid, inner ear, fetal cochlea and kidney, but expression is drastically reduced in thyroid carcinomas. Pendrin functions as a transporter of chloride and iodide, but not sulfate, in these tissues. It is an apical anion transporter in intercalated cells of proximal tubule and cortical collecting ducts, which mediate renal bicarbonate secretion and Cl^-/OH^- , $\text{Cl}^-/\text{HCO}_3^-$ and $\text{Cl}^-/\text{formate}$ exchange in kidney. Pendrin is expressed throughout the endolymphatic duct and sac in distinct areas of the utricle and saccule and in the external sulcus region within the cochlea, where it plays a role in the development of ion gradients.

REFERENCES

1. Everett, L.A., et al. 1997. Pendred syndrome is caused by mutations in a putative sulphate transporter gene (PDS). *Nat. Genet.* 17: 411-422.
2. Coyle, B., et al. 1998. Molecular analysis of the PDS gene in Pendred syndrome. *Hum. Mol. Genet.* 7: 1105-1112.
3. Everett, L.A., et al. 1999. Expression pattern of the mouse ortholog of the Pendred's syndrome gene (PDS) suggests a key role for Pendrin in the inner ear. *Proc. Natl. Acad. Sci. USA* 96: 9727-9732.
4. Scott, D.A., et al. 1999. The Pendred syndrome gene encodes a chloride-iodide transport protein. *Nat. Genet.* 21: 440-443.
5. Scott, D.A., et al. 2000. Human Pendrin expressed in *Xenopus laevis* oocytes mediates chloride/formate exchange. *Am. J. Physiol. Cell Physiol.* 278: C207-C211.
6. Bidart, J.M., et al. 2000. Expression of Pendrin and the Pendred syndrome (PDS) gene in human thyroid tissues. *J. Clin. Endocrinol. Metab.* 85: 2028-2033.

CHROMOSOMAL LOCATION

Genetic locus: SLC26A4 (human) mapping to 7q22.3.

SOURCE

Pendrin (G-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of Pendrin of human origin.

PRODUCT

Each vial contains 200 μg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-23779 P, (100 μg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

RESEARCH USE

For research use only, not for use in diagnostic procedures.

APPLICATIONS

Pendrin (G-19) is recommended for detection of Pendrin of human and, to a lesser extent, rat origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μg per 100-500 μg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Pendrin siRNA (h): sc-44009, Pendrin shRNA Plasmid (h): sc-44009-SH and Pendrin shRNA (h) Lentiviral Particles: sc-44009-V.

Molecular Weight of non-glycosylated Pendrin: 85 kDa.

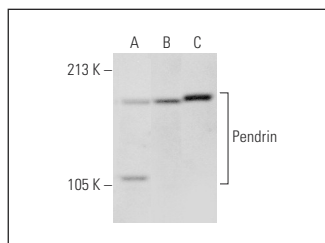
Molecular Weight of glycosylated Pendrin: 110-140 kDa.

Positive Controls: Hep G2 cell lysate: sc-2227, HEK293 whole cell lysate: sc-45136 or COLO 205 whole cell lysate: sc-364177.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

DATA



Pendrin (G-19): sc-23779. Western blot analysis of Pendrin expression in HEK293 (A), Hep G2 (B) and COLO 205 (C) whole cell lysates.

SELECT PRODUCT CITATIONS

1. Purkerson, J.M., et al. 2010. Adaptation to metabolic acidosis and its recovery are associated with changes in anion exchanger distribution and expression in the cortical collecting duct. *Kidney Int.* 78: 993-1005.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.