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GLE1 Rabbit pAb

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Catalog # AP55147

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q53GS7
Reactivity	Rat
Predicted	Pig, Dog, Human, Mouse, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	79836
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human GLE1
Epitope Specificity	611-698/698
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Nucleus. Cytoplasm. Shuttles between the nucleus and the cytoplasm. Shuttling is essential for its mRNA export function and Cytoplasm. Nucleus > nuclear pore complex. Shuttles between the nucleus and the cytoplasm. In the nucleus, isoform 1 localizes to the nuclear pore complex and nuclear envelope. Shuttling is essential for its mRNA export function.
DISEASE	Defects in GLE1 are the cause of lethal congenital contracture syndrome type 1 (LCCS1) [MIM:253310]; also known as multiple contracture syndrome type Finnish. LCCS is an autosomal recessive disorder characterized by early fetal hydrops and akinesia, micrognatia, pulmonary hypoplasia, pterygia, multiple joint contractures, specific neuropathology with degeneration of anterior horn neurons and extreme skeletal muscle atrophy. LCCS1 leads to prenatal death. Defects in GLE1 are the cause of lethal arthrogryposis with anterior horn cell disease (LAAHD) [MIM:611890]. LAAHD is characterized by fetal akinesia, arthrogryposis and motor neuron loss. LAADH fetus often survive delivery, but die early as a result of respiratory failure. Neuropathological findings resemble those of LCCS1, but are less severe.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Protein transport across the nucleus is a selective, multi-step process involving several cytoplasmic factors that mediate protein passage through the nuclear pore complex (NPC). Gle1, also known as GLE1L, is a 698 amino acid protein that localizes to both the nucleus and the cytoplasm and belongs to the Gle1 family. Expressed as two alternatively spliced isoforms, Gle1 associates with the NPC and is required for the transport of poly(A)-containing mRNAs from the nucleus to the cytoplasm. Defects in the gene encoding Gle1 are the cause of lethal congenital contracture syndrome type 1 (LCCS1) and lethal arthrogryposis with anterior horn cell disease (LAAHD), the former of which is characterized by early fetal hydrops and akinesia, micrognatia, pulmonary hypoplasia, pterygia and prenatal death, while the latter is associated with respiratory failure.

Additional Information

Gene ID	2733
Other Names	mRNA export factor GLE1, hGLE1, GLE1 RNA export mediator {ECO:0000303|PubMed:18204449, ECO:0000312|HGNC:HGNC:4315}, GLE1-like protein, Nucleoporin GLE1, GLE1, GLE1L
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	GLE1
Synonyms	GLE1L
Function	Required for the export of mRNAs containing poly(A) tails from the nucleus into the cytoplasm. May be involved in the terminal step of the mRNA transport through the nuclear pore complex (NPC).
Cellular Location	Nucleus. Cytoplasm. Note=Shuttles between the nucleus and the cytoplasm (PubMed:12668658). Shuttling is essential for its mRNA export function (PubMed:12668658).

Background

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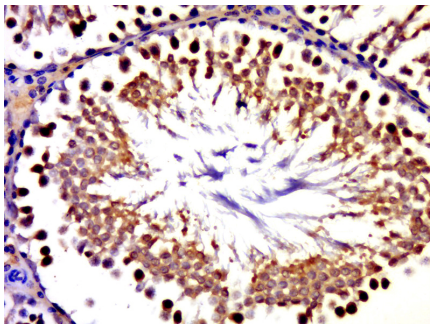
Images



Paraformaldehyde-fixed, paraffin embedded (rat testis); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (GLE1) Polyclonal Antibody, Unconjugated (AP55147) at 1:5000 overnight at 4°C, followed by a conjugated secondary (sp-0023) for 20 minutes and DAB staining.