

ETN3

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Abstract

Endothelin is the most powerful vasoconstrictor. It exists not only in the vascular endothelium, but also in various tissues and cells. It is an important factor in regulating cardiovascular function and maintains basal vascular tone and heart, and plays an important role in the vascular system homeostasis. The endothelin system includes three peptide ligands (EDN1, EDN2, and EDN3) and two G protein-coupled receptors (EDTRA, EDTRB).

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EDN3 interacts with endothelin receptor B(EDNRB) on the surface of cells. Throughout embryonic development EDN3 takes part in neural crest cells that migrate from the developing spinal cord to specific regions in the embryo where they give rise to many different types of cells. EDN3 and EDN3R are necessary for the formation of nerves in the large intestine (enteric nerves) and melanocytes (produce melanin). Mutations in the EDN3 gene is linked with Waardenburg syndrome type IV that is characterized by changes in skin, hair and eye coloring. Mutations in the EDN3 gene is linked with Hirschsprung disease that causes severe constipation or intestinal blockage.