Deleted in Colorectal Carcinoma

Alternative Names
DCC
Colorectal Cancer-Related Chromosome Sequence 18
CRC18
CRCR1

Record Category
Gene locus

WHO-ICD
N/A to gene loci

Incidence per 100,000 Live Births
N/A to gene loci

OMIM Number
120470

Mode of Inheritance
N/A to gene loci

Gene Map Locus
18q21.2

Description
The DCC gene encodes a transmembrane protein that functions as a receptor for netrin-1, a chemoattractant for commissural axons. Hence, in the developing nervous system, upon binding to its ligand, the netrin receptor mediates the guidance of axons in neuronal growth cones towards netrin-1. It may also associate with the UNC5 protein to trigger axon repulsion. By carrying out its functions, DCC plays a key role in the development of nervous system lateralization. DCC can also act as a dependence receptor which induces cell apoptosis in the absence of its ligand. It thus functions as a tumor suppressor gene.

Mutations in the gene have been associated with Somatic Colorectal Cancer and Somatic Esophageal Carcinoma. DCC mutations have also been implicated in the neurological disorders Familial Horizontal Gaze Palsy, with Progressive Scoliosis, 2 (HGPPS2) and Mirror Movements 1 (MRMV1).

Molecular Genetics
The DCC gene, located on the long arm of chromosome 18, spans a length of 1195 kb of DNA. Its coding sequence is spread across 32 exons and it encodes a 158.4 kDa protein product composed of 1447 amino acids. The gene is expressed in a wide variety of cells in the human body, including the intestine, brain, eye and testis. At least 11 heterozygous mutations in the DCC gene have been implicated in mirror movements 1, including missense variants, deletions and premature terminations.

Epidemiology in the Arab World
Saudi Arabia
Monies et al. (2017) illustrated the findings of 1000 diagnostic panels and exomes carried out at a next generation sequencing lab in Saudi Arabia. One patient, a 29-year-old female, suffered from slowly progressive adult-onset ataxia. She did not have any cognitive dysfunction and a nerve conduction study was indicative of sensory neuropathy. She also reported a family history of this phenotype. Using a multigene panel for neurological disorders, a heterozygous mutation (c.1423C>T, p.R475X) was identified in exon 9 of the patient's DCC gene, associated with Mirror Movements 1. Given the atypical presentation of the patient, this case helped in the phenotypic expansion of the disorder.

References
Monies D, Abouelhoda M, AlSayed M, Alhassnan Z, Alotaibi M, Kayyali H, Al-Owain M, Shah A, Rahbeeni Z, Al-Muhaizea MA, Alzaidan HI, Cupler E, Bohlega S, Faqeih E, Faden M, Alyounes B, Jaroudi D, Goljan E, Elbardisy H, Akilan A,

Albar R, Aldhalaan H, Gulab S, Chedrawi A, Al Saud BK, Kurdi W, Makhseed N, Alqasim T, El Khashab HY, Al-Mousa H, Alhashem A, Kanaan I, Algoufi T, Alsaleem K, Basha TA, Al-Murshedi F, Khan S, Al-Kindy A, Alnemer M, Al-Hajjar S, Alyamani S, Aldhekri H, Al-Mehaidib A, Arnaout R, Dabbagh O, Shagrani M, Broering D, Tulbah M, Alqassmi A, Almugbel M, AlQuaiz M, Alsaman A, Al-Thihli K, Sulaiman RA, Al-Dekhail W, Alsaegh A, Bashiri FA, Qari A, Alhomadi S, Alkuraya H, Alsebayel M, Hamad MH, Szonyi L, Abaalkhail F, Al-Mayouf SM, Almojalli H, Alqadi KS, Elsiesy H, Shuaib TM, Seidahmed MZ, Abosoudah I, Akleh H, AlGhonaium A, Alkharfy TM, Al Mutairi F, Eyaid W, Alshanbary A, Sheikh FR, Alsohaibani FI, Alsonbul A, Al Tala S, Balkhy S, Bassiouni R, Alenizi AS, Hussein MH, Hassan S, Khalil M, Tabarki B, Alshahwan S, Oshi A, Sabr Y, Alsaadoun S, Salih MA, Mohamed S, Sultana H, Tamim A, El-Haj M, Alshahrani S, Bubshait DK, Alfadhel M, Faquih T, El-Kalioby M, Subhani S, Shah Z, Moghrabi N, Meyer BF, Alkuraya FS. The landscape of genetic diseases in Saudi Arabia based on the first 1000 diagnostic panels and exomes. Hum Genet. 2017 Aug;136(8):921-939. PMID: 28600779.

Related CTGA Records

Mirror Movements 1 (OMIM 157600)

External Links

http://www.genecards.org/cgi-bin/carddisp.pl?gene=DCC

Contributors

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