

## DICER1 gene

dicer 1, ribonuclease III

### Normal Function

The *DICER1* gene provides instructions for making a protein that plays a role in regulating the activity (expression) of other genes. The Dicer protein aids in the production of a molecule called microRNA (miRNA). MicroRNAs are short lengths of RNA, a chemical cousin of DNA. Dicer cuts (cleaves) precursor RNA molecules to produce miRNA.

MicroRNAs control gene expression by blocking the process of protein production. In the first step of making a protein from a gene, another type of RNA called messenger RNA (mRNA) is formed and acts as the blueprint for protein production. MicroRNAs attach to specific mRNA molecules and stop the process by which protein is made. Sometimes, miRNAs break down the mRNA, which also blocks protein production. Through this role in regulating the expression of genes, Dicer is involved in many processes, including cell growth and division (proliferation) and the maturation of cells to take on specialized functions (differentiation).

### Health Conditions Related to Genetic Changes

#### DICER1 syndrome

Mutations in the *DICER1* gene cause *DICER1* syndrome. People with this condition have an increased risk of developing many types of tumors, particularly certain tumors of the lungs (pleuropulmonary blastoma); kidneys (cystic nephroma); ovaries (Sertoli-Leydig tumors); and thyroid, a butterfly-shaped gland in the lower neck (multinodular goiter). Most of these mutations lead to an abnormally short Dicer protein that is likely unable to produce miRNA. Without regulation by miRNA, genes are expressed abnormally, which could cause cells to grow and divide uncontrollably and lead to tumor formation.

### Other Names for This Gene

- DCR1
- Dicer
- dicer 1 ribonuclease III

- dicer 1, double-stranded RNA-specific endoribonuclease
- dicer 1, ribonuclease type III
- Dicer1, Dcr-1 homolog
- DICER\_HUMAN
- endoribonuclease Dicer
- helicase MOI
- helicase with RNase motif
- helicase-moi
- HERNA
- K12H4.8-LIKE
- KIAA0928
- MNG1

## **Additional Information & Resources**

### Tests Listed in the Genetic Testing Registry

- Tests of DICER1 ([https://www.ncbi.nlm.nih.gov/gtr/all/tests/?term=23405\[geneid\]](https://www.ncbi.nlm.nih.gov/gtr/all/tests/?term=23405[geneid]))

### Scientific Articles on PubMed

- PubMed (<https://pubmed.ncbi.nlm.nih.gov/?term=%28DICER1%5BTIAB%5D%29+AND+%28%28Genes%5BMH%5D%29+OR+%28Genetic+Phenomena%5BMH%5D%29%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+720+days%22%5Bdp%5D>)

### Catalog of Genes and Diseases from OMIM

- DICER 1, RIBONUCLEASE III; DICER1 (<https://omim.org/entry/606241>)

### Gene and Variant Databases

- NCBI Gene (<https://www.ncbi.nlm.nih.gov/gene/23405>)
- ClinVar ([https://www.ncbi.nlm.nih.gov/clinvar?term=DICER1\[gene\]](https://www.ncbi.nlm.nih.gov/clinvar?term=DICER1[gene]))

## **References**

- Bahubeshi A, Bal N, Rio Frio T, Hamel N, Pouchet C, Yilmaz A, Bouron-DalSoglio D, Williams GM, Tischkowitz M, Priest JR, Foulkes WD. Germline DICER1 mutations and familial cystic nephroma. J Med Genet. 2010 Dec;47(12):863-6. doi:10.1136/jmg.

2010.081216. Epub 2010 Oct 29. Citation on PubMed (<https://pubmed.ncbi.nlm.nih.gov/21036787>)

- Carthew RW, Sontheimer EJ. Origins and Mechanisms of miRNAs and siRNAs. *Cell*. 2009 Feb 20;136(4):642-55. doi: 10.1016/j.cell.2009.01.035. Citation on PubMed (<https://pubmed.ncbi.nlm.nih.gov/19239886>) or Free article on PubMed Central (<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2675692/>)
- Ghildiyal M, Zamore PD. Small silencing RNAs: an expanding universe. *Nat Rev Genet*. 2009 Feb;10(2):94-108. doi: 10.1038/nrg2504. Citation on PubMed (<https://pubmed.ncbi.nlm.nih.gov/19148191>) or Free article on PubMed Central (<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC2724769/>)
- Rio Frio T, Bahubeshi A, Kanellopoulou C, Hamel N, Niedziela M, Sabbaghian N, Pouchet C, Gilbert L, O'Brien PK, Serfas K, Broderick P, Houlston RS, Lesueur F, Bonora E, Muljo S, Schimke RN, Bouron-Dal Soglio D, Arseneau J, Schultz KA, Priest JR, Nguyen VH, Harach HR, Livingston DM, Foulkes WD, Tischkowitz M. DICER1 mutations in familial multinodular goiter with and without ovarian Sertoli-Leydig cell tumors. *JAMA*. 2011 Jan 5;305(1):68-77. doi: 10.1001/jama.2010.1910. Citation on PubMed (<https://pubmed.ncbi.nlm.nih.gov/21205968>) or Free article on PubMed Central (<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3406486/>)
- Schultz KA, Pacheco MC, Yang J, Williams GM, Messinger Y, Hill DA, Dehner LP, Priest JR. Ovarian sex cord-stromal tumors, pleuropulmonary blastoma and DICER1 mutations: a report from the International Pleuropulmonary Blastoma Registry. *Gynecol Oncol*. 2011 Aug;122(2):246-50. doi: 10.1016/j.ygyno.2011.03.024. Epub 2011 Apr 17. Citation on PubMed (<https://pubmed.ncbi.nlm.nih.gov/21501861>) or Free article on PubMed Central (<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3138876/>)
- Slade I, Bacchelli C, Davies H, Murray A, Abbaszadeh F, Hanks S, Barfoot R, Burke A, Chisholm J, Hewitt M, Jenkinson H, King D, Morland B, Pizer B, Prescott K, Saggat A, Side L, Traunecker H, Vaidya S, Ward P, Futreal PA, Vujanic G, Nicholson AG, Sebire N, Turnbull C, Priest JR, Pritchard-Jones K, Houlston R, Stiller C, Stratton MR, Douglas J, Rahman N. DICER1 syndrome: clarifying the diagnosis, clinical features and management implications of a pleiotropic tumour predisposition syndrome. *J Med Genet*. 2011 Apr;48(4):273-8. doi:10.1136/jmg.2010.083790. Epub 2011 Jan 25. Citation on PubMed (<https://pubmed.ncbi.nlm.nih.gov/21266384>)

## Genomic Location

The *DICER1* gene is found on chromosome 14 (<https://medlineplus.gov/genetics/chromosome/14/>).

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