



# Huntington Disease Genemer™ Control DNA\*

\*Specific control DNA for use with Gene Link Genemer™ & GeneProber™ product lines

Catalog No. 40-2025-XX

**For research use only. Not for use in diagnostic procedures for clinical purposes**

Product	Catalog Number	Unit Size
<input type="checkbox"/> GLHD ~18 CAG repeat Genemer Control DNA	40-2025-01	500 ng
<input type="checkbox"/> GLHD ~34 CAG repeat Genemer Control DNA	40-2025-02	500 ng
<input type="checkbox"/> GLHD ~44 CAG repeat Genemer Control DNA	40-2025-03	500 ng
<input type="checkbox"/> GLHD ~89 CAG repeat Genemer Control DNA	40-2025-04	500 ng
<input type="checkbox"/> GLHD ~116 CAG repeat Genemer Control DNA	40-2025-06	500 ng
<input type="checkbox"/> GLHD ~134 CAG repeat Genemer Control DNA	40-2025-05	500 ng

### Background

Huntington disease (HD) is an autosomal dominant, progressive neurodegenerative disorder with a prevalence rate of about 5-10 affected persons per 100,000 in most western populations. The disorder presents with motor impairment, cognitive deterioration and psychiatric symptoms.

HD is caused by a CAG trinucleotide expansion within the first exon of the *IT15* gene on chromosome 4p16. The expanded CAG repeats are translated into a polyglutamine tract in the Huntington protein, which is believed to cause a dominant gain of function, leading to neuronal dysfunction and neurodegeneration.

The number of CAG repeats correlates inversely with the age of onset of symptoms. The American College of Medical Genetics/American Society of Human Genetics/Huntington Disease Genetics Testing Working Group divided the genotype/phenotype correlation in the following four categories for CAG repeat lengths: normal allele, ≤26 CAG repeats, generating a normal phenotype; intermediate allele, 27-35 CAG repeats, mutable normal allele generating a normal phenotype; HD allele with reduced penetrance, 36-39 CAG repeats, generating a normal or HD phenotype; HD allele, ≥40 CAG repeats, generating a HD phenotype.

The CAG trinucleotide expansion is unstable and can lengthen during transmission from parents to offspring. Thus the age of onset can decrease from one generation to the next, a phenomenon known as anticipation. HD anticipation is more intense in paternal transmission.

### Genotyping

Polymerase Chain Reaction (PCR) is the most common method used to estimate the number of CAG repeats. Since the CAG repeats in the HD gene are immediately 5' of a CCG repeat which is also polymorphic in

length, the PCR product of this primer pair excludes the known adjacent polymorphic CCG repeat that can contribute to an inaccurate determination of HD gene CAG repeat sizes in individuals who may have an HD gene CAG repeat allele close to the normal/affected boundary.

### Material Supplied

A tube containing 500 ng of lyophilized control DNA segment. The above control DNA is an ideal genotyping template for optimizing and performing control amplification with unknown DNA. The size of the triple repeats has been determined by sequencing and gel electrophoresis. The stability of size repeats upon cloning and amplification has NOT been determined. Thus, the size should be considered approximate and there is no claim for each fragment to contain the exact number of triple repeats.

The quantity supplied is sufficient for 1000 regular 50µl PCR\*\* reaction.

### Reconstitution

**Stock Solution:** Add 100µl sterile water to the tube containing the lyophilized DNA to yield a solution of 5 ng/µl.

**Working Solution:** Dilute 1:10 an aliquot of the stock solution

**Usage:** Initially use 1µl each of the stock and working template solution for amplification and optimization of the reaction. Based on the results, use 1µl of template at the lowest concentration.

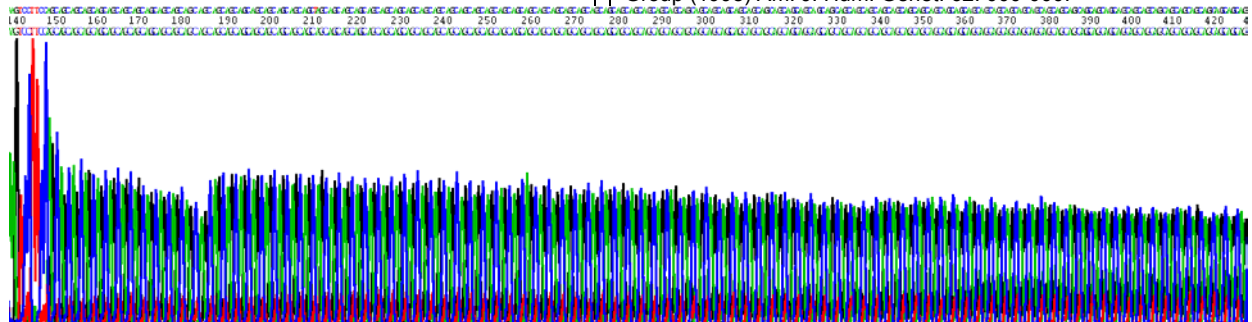
### Protocol for PCR Analysis of Triple Repeat Size

Follow protocol supplied with the appropriate Genemer™ or GeneProber™ product

### References:

Kremer, B. et al. (1993) N. Engl. J. Med. 330: 1401-1406.

The American College of Medical Genetics/American Society of Human Genetics Huntington Disease Genetic Testing Working Group (1998) Am. J. Hum. Genet. 62: 000-000.



## Huntington Disease Product Ordering Information

Product	Size	Catalog No.	Price, \$
Huntington Disease Genemer™ Primer pair for amplification of CAG triple repeat spanning region. The quantity supplied is sufficient for 400 regular 50 µl PCR reactions.	10 nmols	40-2025-10	\$100.00
Huntington Disease GeneProber™ GLHD Probe unlabeled	500 ng	40-2025-40	\$350.00
Huntington Disease CTG triple repeat spanning region unlabeled probe for radioactive labeling and Southern blot detection. Suitable for random primer labeling.	110 µl	40-2025-41	\$400.00
Huntington Disease GeneProber™ GLHD Probe Digoxigenin labeled	12 µl	40-2025-31	\$400.00
Huntington Disease CAG repeat spanning region digoxigenin labeled probe for non-radioactive detection Southern blot.	5 blots	40-2025-32	\$650.00
Huntington Disease PCRProber™ AP labeled probe Alkaline phosphatase labeled probe	5 blots	40-2025-32	\$650.00
Huntington Disease PCRProber™ Kit. Kit for performing non-radioactive PCR amplification based detection. 5 blots (50 rxns)			
<b>Genemer™ control DNA</b> Cloned fragment of the mutation region of a particular gene. These control DNA's are ideal genotyping templates for optimizing and performing control amplification with unknown DNA. The size of the triple repeats has been determined by sequencing and gel electrophoresis. The stability of size repeats upon cloning and amplification has NOT been determined. Thus, the size should be considered approximate and there is no claim for each fragment to contain the exact number of triple repeats.			
GLHD 18 ~CAG repeat Genemer Control DNA	500 ng	40-2025-01	175.00
GLHD 34 ~CAG repeat Genemer Control DNA	500 ng	40-2025-02	175.00
GLHD 44 ~CAG repeat Genemer Control DNA	500 ng	40-2025-03	175.00
GLHD 89 ~CAG repeat Genemer Control DNA	500 ng	40-2025-04	175.00
GLHD 116 ~CAG repeat Genemer Control DNA	500 ng	40-2025-06	175.00
GLHD 134 ~CAG repeat Genemer Control DNA	500 ng	40-2025-05	175.00

Please visit [www.genelink.com](http://www.genelink.com) for other Genemer™ control DNA not listed here

### Genemer™ Control DNA (Selected List)

Product	Size	Catalog No.	Price, \$
Fragile X, various CGG triple repeat region control DNA	500 ng	40-2004-XX	175.00
Huntington Disease various CAG triple repeat region control DNA	500 ng	40-2025-XX	175.00
Myotonic Dystrophy various CTG triple repeat region control DNA	500 ng	40-2026-XX	175.00
Friedreich's Ataxia, various GAA triple repeat region control DNA	500 ng	40-2027-XX	175.00

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### Genemer™ (Selected List)

Product	Size	Catalog No.	Price, \$
Fragile X (spanning triple repeat region)	10nmols	40-2004-10	100.00
Huntington Disease (spanning triple repeat region)	10nmols	40-2025-10	100.00
Myotonic Dystrophy (spanning triple repeat region)	10nmols	40-2026-10	100.00
Friedreich's Ataxia	10nmols	40-2027-10	100.00
Factor V	10nmols	40-2035-10	100.00
Factor VIII (Hemophilia)	10nmols	40-2036-10	100.00
STS (Steroid Sulfatase)	10nmols	40-2023-10	100.00
HGH (Human Growth Hormone)	10nmols	40-2024-10	100.00
Sickle Cell	10nmols	40-2001-10	100.00
RhD (Rh D gene exon 10 specific)	10nmols	40-2002-10	100.00
Rh EeCc (Rh Ee and Cc exon 7 specific)	10nmols	40-2003-10	100.00
Gaucher (various mutations)	10nmols	40-2047-10	100.00
Cystic Fibrosis (various mutations)	10nmols	40-2029-10	100.00
SRY (sex determining region on Y)	10nmols	40-2020-10	100.00
X alphoid repeat	10nmols	40-2021-10	100.00
Y alphoid repeat	10nmols	40-2022-10	100.00

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\*\*The polymerase chain reaction (PCR) process is covered by patents owned by Hoffmann-La Roche. A license to perform is automatically granted by the use of authorized reagents.

Prices subject to change without notice

All Gene Link products are for research use only



140 Old Saw Mill River Road Hawthorne, NY 10532  
Tel: 914.769.1192 [www.genelink.com](http://www.genelink.com) Fax: 914.769.1193