



Product Sheet

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.

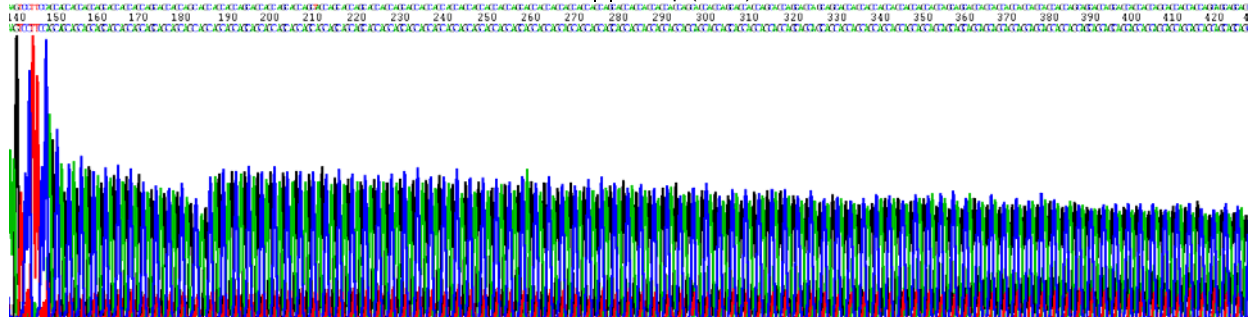
Protocol for PCR Analysis of Triple Repeat Size

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.



GeneProber™ Product Ordering Information

The GeneProber™ product line is based on the chemiluminescent Southern blot detection method. Gene Link's non-radioactive detection systems for genotyping of triple repeat disorders are rapid, reliable and as sensitive as the ³²P labeled southern blots. No more decayed probes and radioactive exposure. Kits are available for reliable genotyping of the fragile X, myotonic dystrophy and other triple repeat mutation group disorders.

Unlabeled GeneProber™ probes are also available for radio labeling and radioactive based detection. Gene Link strongly recommends the use of non-radioactive gene detection systems. Consider switching to Gene Link's product line of non-radioactive detection systems

Product	Unit Size	Catalog No.
Fragile X GeneProber™ GLFX1 Probe unlabeled	500 ng	40-2004-40
Fragile X GeneProber™ GLFXDig1 Probe Digoxigenin labeled	110 µL	40-2004-41
Huntington's Disease GeneProber™ GLHD14 Probe unlabeled	500 ng	40-2025-40
Huntington's Disease GeneProber™ GLHDDig2X Probe Digoxigenin labeled	110 µL	40-2025-41
Myotonic Dystrophy GeneProber™ GLDM1 Probe unlabeled	500 ng	40-2026-40
Myotonic Dystrophy GeneProber™ GLDMDig2 Probe Digoxigenin labeled	110 µL	40-2026-41
Friedreich's Ataxia GeneProber™ GLFRDA21 Probe unlabeled	500 ng	40-2027-40
Friedreich's Ataxia GeneProber™ GLFRDADig21 Probe Digoxigenin labeled	110 µL	40-2027-41

GScan™ Products Product Ordering Information

Gene Link's GScan™ gene detection products are safe, convenient and sensitive, and afford automated compilation of data. The kits contain optimized PCR amplification reagents and a wide array of fluorescent-labeled primers for genotyping after PCR using fluorescent genetic analyzer instrument(s). Included in these kits are ready-to-run control samples of various repeats of the triple repeat disorder kit. These control samples are for calibration with the molecular weight markers for accurate size determination of the amplified fragments.

The GScan™ kits are simple and robust for routine triple-repeat detection of greater than 100 repeats of all triple repeat disorders listed, except Fragile X. The CGG repeat in Fragile X can be detected up to ~50 repeats.

Product	Unit Size	Catalog No.
Fragile X GScan™ Kit for fluorescent detection; 100 reactions kit	1 kit	40-2004-15XX
Fragile X GScan™ Kit for fluorescent detection; 20 reactions kit	1 kit	40-2004-15FMS
Huntington's Disease GScan™ Kit for fluorescent detection; 100 reactions kit	1 kit	40-2025-15XX
Huntington's Disease GScan™ Kit for fluorescent detection; 20 reactions kit	1 kit	40-2025-15FMS
Myotonic Dystrophy GScan™ Kit for fluorescent detection; 100 reactions kit	1 kit	40-2026-15XX
Myotonic Dystrophy GScan™ Kit for fluorescent detection; 20 reactions kit	1 kit	40-2026-15FMS
Friedreich's Ataxia GScan™ Kit for fluorescent detection; 100 reactions kit	1 kit	40-2027-15XX
Friedreich's Ataxia GScan™ Kit for fluorescent detection; 20 reactions kit	1 kit	40-2027-15FMS

All Gene Link products are for research use only

Current pricing are posted at <http://www.genelink.com/>

Huntington's Disease Product Ordering Information

Product	Unit Size	Catalog No.
Huntington's Disease Genemer™ Primer pair Primers 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
Huntington's Disease PCRProber™ AP labeled probe Alkaline phosphatase labeled probe	12 µL	40-2025-31
Huntington's Disease PCRProber™ Kit for chemiluminescent detection Kit for performing PCR amplification and chemiluminescent based detection.	5 blots [50 rxns]	40-2025-32
Huntington's Disease Genemer™ Kit for Radioactive Detection Kit for amplification and radioactive detection of Huntington's Disease CAG triple repeat region amplified PCR products using ³⁵ S or ³² P. 100 Reactions.	1 Kit [100 rxns]	40-2025-20
Huntington's Disease GScan Kit for fluorescent detection Kit for performing fluorescent PCR amplification based detection. Various dye kits. XX=FM for 6-Fam; HX for Hex; TT for Tet; C3 for Cy3 and C5 for Cy5.	1 Kit [100 rxns]	40-2025-15XX

Genemer™ GScan Control DNA Cloned fragment of the mutation region of a particular gene. These control DNAs 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. These control DNAs are sold with the express condition that these NOT be used for exact triple repeat size determination of DNA of unknown genotype. The control DNA should be used for determining the performance of specific Genemer™ and PCRProber™ Gene Link products.

Huntington's Disease 7 ~CAG repeat GScan Genemer Control DNA; HEX labeled	25 µL	40-2025-05HX
Huntington's Disease 18 ~CAG repeat GScan Genemer Control DNA; HEX labeled	25 µL	40-2025-01HX
Huntington's Disease 31 ~CAG repeat GScan Genemer Control DNA; HEX labeled	25 µL	40-2025-07HX
Huntington's Disease 34 ~CAG repeat GScan Genemer Control DNA; HEX labeled	25 µL	40-2025-02HX
Huntington's Disease 37 ~CAG repeat GScan Genemer Control DNA; HEX labeled	25 µL	40-2025-08HX
Huntington's Disease 44 ~CAG repeat GScan Genemer Control DNA	25 µL	40-2025-03HX
Huntington's Disease 49 ~CAG repeat GScan Genemer Control DNA	25 µL	40-2025-09HX
Huntington's Disease 89 ~CAG repeat GScan Genemer Control DNA	25 µL	40-2025-04HX
Huntington's Disease 116 ~CAG repeat GScan Genemer Control DNA	25 µL	40-2025-06HX
Huntington's Disease 134 ~CAG repeat GScan Genemer Control DNA	25 µL	40-2025-61HX
Huntington's Disease 182 ~CAG repeat GScan Genemer Control DNA	25 µL	40-2025-62HX

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Omni-Pure™ DNA & RNA Purification Systems

Product	Catalog No.	Unit Size*(Purifications)
Omni-Pure™ Blood DNA Purification System	40-4010-01	100
Omni-Pure™ Blood DNA Purification System	40-4010-05	500
Omni-Pure™ Blood DNA Purification System	40-4010-10	1000
Omni-Pure™ Tissue DNA Purification System	40-4050-01	100
Omni-Pure™ Tissue DNA Purification System	40-4050-05	500
Omni-Pure™ Tissue DNA Purification System	40-4050-10	1000
Omni-Pure™ Plant DNA Purification System	40-4060-01	100
Omni-Pure™ Plant DNA Purification System	40-4060-05	500
Omni-Pure™ Plant DNA Purification System	40-4060-10	1000
Omni-Pure™ Viral DNA Purification System	40-3720-01	100
Omni-Pure™ Viral DNA Purification System	40-3720-05	500
Omni-Pure™ Microbial DNA Purification System	40-3700-01	100
Omni-Pure™ Microbial DNA Purification System	40-3700-05	500
Omni-Pure™ Viral RNA Purification System	40-3650-01	100
Omni-Pure™ Viral RNA Purification System	40-3650-05	500

*Sample volume for each purification system varies. Each purification yields sufficient quantity for desired applications.

Omni-Clean™ Gel DNA Purification and Concentration Systems

Product	Catalog No.	Unit Size*(Purifications)
Omni-Clean™ Gel DNA Beads Purification System	40-4110-10	100
Omni-Clean™ Gel DNA Beads Purification System	40-4110-50	500
Omni-Clean™ Gel DNA Spin Column Purification System	40-4120-10	100
Omni-Clean™ Gel DNA Spin Column Purification System	40-4120-50	500
Omni-Clean™ DNA Beads Concentration System	40-4130-10	100
Omni-Clean™ DNA Beads Concentration System	40-4130-50	500
Omni-Clean™ DNA Spin Column Concentration System	40-4140-10	100
Omni-Clean™ DNA Spin Column Concentration System	40-4140-50	500

*Sample volume for each purification system varies. Each purification yields sufficient quantity for desired applications.

Omni-Pure™ Plasmid DNA Purification Systems

Product	Catalog No.	Unit Size*(Purifications)
Omni-Pure™ Plasmid DNA Purification System	40-4020-01	100
Omni-Pure™ Plasmid DNA Purification System	40-4020-05	500

*Sample volume for each purification system varies. Each purification yields sufficient quantity for desired applications.

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Southern Blot Buffers & Reagents

Product	Catalog No.	Unit Size
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Agarose Tablets, 0.5 gm each; 100 tablets	40-3011-10	100 tablets
Agarose LE Molecular Biology Grade; 100 g	40-3010-10	100 g
Agarose LE Molecular Biology Grade; 500 g	40-3010-50	500 g
Hybwash A, Hybridization Wash Solution (20X SSC); 200 mL	40-5020-20	200 mL
Hybwash B, Hybridization Wash Solution (10% SDS); 100 mL	40-5021-10	100 mL
TAE Buffer; 50 X Concentrate; 100 mL	40-3007-01	100 mL
TAE Buffer; 50 X Concentrate; 1 L	40-3007-10	1 L
TBE Buffer; 5 X Concentrate; 1 L	40-3008-10	1 L
Buffer M 10X (Maleic Acid buffer); 100 mL	40-5025-10	100 mL
10% Blocking solution; 100 mL	40-5026-10	100 mL
Loading Buffer 2X BPB/XC Denaturing for Sequencing; 1 mL	40-5027-10	1 mL
10x AP Detection buffer (alkaline phosphatase detection buffer); 100 mL	40-5031-10	100 mL
Lumisol™ I Hybridization Solution; contains formamide; 200 mL	40-5022-20	200 mL
Lumisol™ II Hybridization Solution; for non-toxic hybridizations; 200 mL	40-5023-20	200 mL
Lumisol™ III Hybridization Solution; for oligo probes; 200 mL	40-5024-20	200 mL
CDP-Star® Substrate; Ready-to-Use 0.25 mM in spray bottle; 10 mL	40-5010-10	10 mL

Loading Buffers

Product	Catalog No.	Size
Gel Loading Buffer 5X BPB/XC non-denaturing; 1 mL	40-3002-10	1 mL
Gel Loading Buffer 5X BPB/XC non-denaturing; 15 mL	40-3002-15	15 mL
Gel Loading Buffer 10X BPB/XC non-denaturing; 1 mL	40-3003-10	1 mL
Gel Loading Buffer 10X BPB/XC non-denaturing; 15 mL	40-3003-15	15 mL
Gel Loading Buffer 5X Orange G/XC non-denaturing; 1 mL	40-3004-10	1 mL
Gel Loading Buffer 5X Orange G/XC non-denaturing; 15 mL	40-3004-15	15 mL
Gel Loading Buffer 2X BPB/XC Denaturing for Sequencing; 1 mL	40-5027-10	1 mL
Gel Loading Buffer 2X BPB/XC Denaturing for Sequencing; 15 mL	40-5027-15	15 mL
DNA SDS Gel Loading Buffer 5X BPB/XC DNA binding protein denaturing buffer ; 1 mL	40-5028-10	1 mL
DNA SDS Gel Loading Buffer 5X BPB/XC DNA binding protein denaturing buffer; 15 mL	40-5028-15	15 mL
RNA Gel Loading Buffer 2X BPB/XC with ethidium bromide; 1 mL	40-5029-10	1 mL
RNA Gel Loading Buffer 2X BPB/XC with ethidium bromide; 15 mL	40-5029-15	15 mL
RNA Gel Loading Buffer 2X BPB/XC without ethidium bromide ; 1 mL	40-5030-10	1 mL
RNA Gel Loading Buffer 2X BPB/XC without ethidium bromide; 15 mL	40-5030-15	15 mL

Omni-Marker™

Product	Catalog No.	Size*
Omni-Marker™ Universal unlabeled; 1 mL	40-3005-10	1 mL
Omni-Marker™ Low unlabeled; 1 mL	40-3006-10	1 mL
Omni-Marker™ GScan™-2 Tamra labeled 50 bp - 600 bp; 500 µL	40-3062-05	500 µL

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