

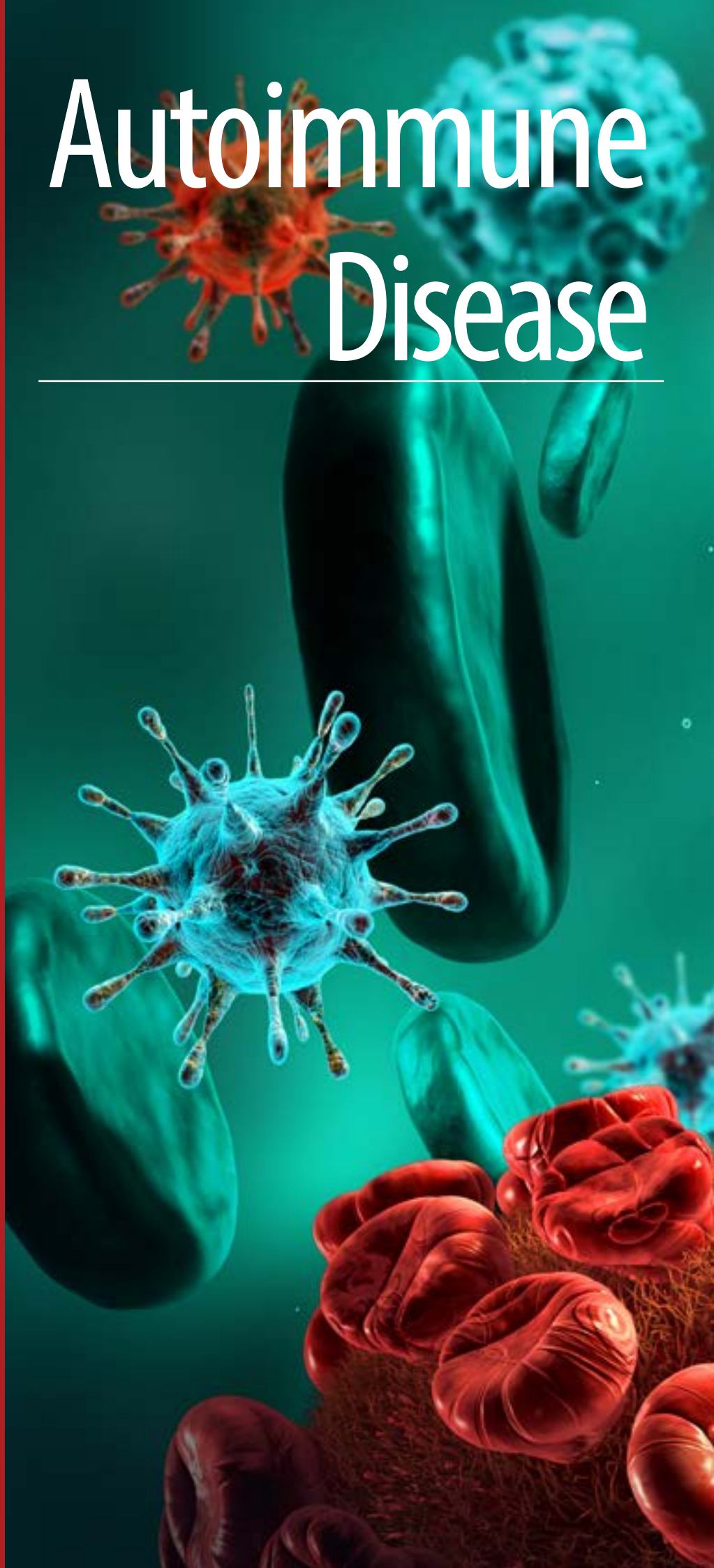


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# Autoimmune Disease

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## Amphiphysin protein

Also known as: AMPH protein, AMPH1 protein, Amphiphysin protein, Stiff-Mann syndrome with breast cancer 128kD autoantigen. Amphiphysin is a protein associated with the cytoplasmic surface of synaptic vesicles. Amphiphysin is a brain-enriched protein with an N-terminal lipid interaction, dimerisation and membrane bending BAR domain, a middle clathrin and adaptor binding domain and a C-terminal SH3 domain. In the brain, its primary function is thought to be the recruitment of dynamin to sites of clathrin-mediated endocytosis.

### Amphiphysin protein in Disease

Antibodies against AMPH are detected in patients with Stiff Person Syndrome (SPS), a rare disease of the central nervous system characterized by progressive rigidity of the body musculature with superimposed painful spasms. A subset of patients with SPS who were also affected by breast cancer are positive for autoantibodies against this protein.

Catalog No.	Amino Acids	Expression System	Purity
80-1531	1-695 aa	E.coli	>90% pure

## DPYSL5 protein

Also known as: CRIM3 protein, BMP binding endothelial regulator protein, BMP-binding endothelial regulator precursor protein, BMPER protein, bone morphogenetic protein-binding endothelial cell precursor-derived regulator protein, crossveinless 2 protein, crossveinless-2 protein, CV-2 protein, CV2 protein, Q8N8U9 protein

DPYSL5 is a secreted protein that interacts with, and inhibits bone morphogenetic protein (BMP) function. It has been shown to inhibit BMP2- and BMP4-dependent osteoblast differentiation and BMP-dependent differentiation of the chondrogenic cells. DPYSL5 is a member of the collapsin response mediator protein family (CRMPs) which are involved in an autoimmune disease associated with small cell lung carcinoma. Members of the CRMP family, such as DPYSL5, are believed to play a role in growth cone guidance during neural development.

### DPYSL5 protein in Disease

Defects in DPYSL5 are the cause of diaphanospondylodysostosis (DSD), a rare, recessively inherited, perinatal lethal skeletal disorder, the characteristics of which include a small chest, abnormal vertebral segmentation, as well as craniofacial abnormalities.

Catalog No.	Amino Acids	Expression System	Purity
80-1532	1-564 aa	E.coli	>85% pure

## dsDNA

Also known as: Double Stranded DNA, purified dsDNA

Most DNA molecules are actually two polymer strands, bound together in a helical fashion by noncovalent bonds; this double stranded structure (dsDNA) is maintained largely by the intrastrand base stacking interactions, which are strongest for G, C stacks. The two strands can come apart – a process known as melting – to form two single-stranded DNA molecules (ssDNA) molecules.

### dsDNA in Disease

Anti-dsDNA antibodies target double stranded DNA. They are highly diagnostic of Systemic Lupus Erythematosus (SLE) and are implicated in the pathogenesis of Lupus Nephritis.

Catalog No.	Expression System
30-1805	E.coli

## GARS Protein

Also known as: Glycyl-tRNA synthetase protein, GlyRS protein, EJ protein

GARS is found in all cell types and plays an important role in the production (synthesis) of proteins. The GARS gene provides instructions for making an enzyme called glycyl-tRNA synthetase which plays a role in adding the amino acid glycine at the appropriate point on an amino acid chain.

### GARS protein in Disease

Charcot-Marie-Tooth disease is caused by mutations in the GARS gene. It is thought that the mutations reduce the activity of glycyl-tRNA synthetase. Scientists suggest that nerve cells may be especially sensitive to a decrease in the activity of this enzyme. In particular, a reduction in glycyl-tRNA synthetase activity may impair the ability of specialized outgrowths from nerve cells (axons) to transmit nerve impulses.

Distal Hereditary Motor Neuropathy, Type V is also caused by mutations in the GARS gene. Similar to Charcot-Marie-Tooth disease, a reduction in glycyl-tRNA synthetase activity may impair transmission of nerve impulses.

Catalog No.	Amino Acids	Expression System	Purity
80-1374	55-739 with 6x His tag	E.coli	>85% pure

## IARS Protein

Also known as: Isoleucyl-tRNA Synthetase protein, IleRS protein, OJ protein

IARS has a central role in linking amino acids with nucleotide triplets contained in tRNAs. Aminoacyl-tRNA synthetases catalyze the aminoacylation of tRNA by their cognate amino acid and are thought to be among the first proteins that appeared in evolution.

### IARS protein in Disease

Isoleucine-tRNA synthetase belongs to the class-I aminoacyl-tRNA synthetase family and has been identified as a target of auto-antibodies in the autoimmune disease Polymyositis/Dermatomyositis. Two alternatively spliced variants have been isolated that represent alternate 5' UTRs.

Catalog No.	Amino Acids	Expression System	Purity
80-1377	1-1262 with 6x His tag	Baculovirus	>75% pure

## JO1 Protein

Also known as: Histidyl-tRNA Synthetase protein, HisRS protein, JO-1 protein, JO 1 protein, HARS protein

JO1 is a cytoplasmic enzyme which belongs to the class II family of aminoacyl-tRNA synthetases. The enzyme is responsible for the synthesis of histidyl-transfer RNA, which is essential for the incorporation of histidine into proteins.

### JO1 protein in Disease

JO1 antibodies are a marker for the disease Polymyositis, and occur most commonly in myositis patients who also have interstitial lung disease. The antibodies occur in up to 50% of patients with interstitial pulmonary fibrosis and symmetrical polyarthritis. JO1 antibodies in patients with polymyositis bind to conformational epitopes of the enzyme protein and inhibit its catalytic activity in vitro.

Catalog No.	Amino Acids	Expression System	Purity
80-1375	1-509 with 6x His tag	E.coli	>90% pure

## PDHE2 Protein

Also known as: Pyruvate Dehydrogenase Complex Component E2 protein, PDH-E2 protein, PDH E2 protein

The pyruvate dehydrogenase complex contributes to transforming pyruvate into acetyl-CoA by a process called pyruvate decarboxylation. Pyruvate dehydrogenase (E1) performs the first two reactions within the pyruvate dehydrogenase complex (PDC): a decarboxylation of substrate 1 (pyruvate) and a reductive acetylation of substrate 2 (lipoic acid). Lipoic acid is covalently bound to dihydrolipoamide acetyltransferase (E2), which is the second catalytic component enzyme of PDC.

### PDHE2 protein in Disease

Pyruvate dehydrogenase is an autoantigen recognized in primary biliary Cirrhosis. These antibodies appear to recognize oxidized protein that has been produced from inflammatory immune responses. It is thought that some of these inflammatory responses could be related to gluten sensitivity. Pyruvate dehydrogenase (PDH) deficiency is a congenital degenerative metabolic disease resulting from a mutation of the pyruvate dehydrogenase complex (PDC) located on the X chromosome. PDH deficiency is a common cause of lactic acidosis in newborns and often presents with severe lethargy, poor feeding, tachypnea, and cases of death have occurred.

Catalog No.	Amino Acids	Expression System	Purity
80-1378	168-313 with 6x His tag and GST tag	E.coli	>90% pure

## POLR3A protein

Also known as: DNA-directed RNA polymerase III largest subunit protein, hRPC155 protein, RNA polymerase III 155 kDa subunit protein, RNA polymerase III subunit C1 protein, RNA polymerase III subunit C160 protein, RNA polymerase III subunit RPC155-D protein, RPC1 protein, RPC155 protein.

POLR3A is the largest and catalytic core component of RNA polymerase III which synthesizes small RNAs, such as 5S rRNA and tRNAs. POLR3A forms the polymerase active center together with the second largest subunit. POLR3A also plays a key role in sensing and limiting infection by intracellular bacteria and DNA viruses and acts as nuclear and cytosolic DNA sensor involved in innate immune response.

### POLR3A protein in Disease

Defects and mutations in POLR3A have been associated with Leukodystrophy, an autosomal recessive neurodegenerative disorder characterized by childhood onset of progressive motor decline manifest as spasticity, ataxia, tremor, and cerebellar signs.

Catalog No.	Amino Acids	Expression System	Purity
80-1533	891-1090 aa	E.coli	>90% pure

## PSIP1 protein & PSIP fragment protein

Also known as: Dense Fine Speckle Protein fragment protein, DSF70 fragment protein, PSIP protein, PC4 and SFRS1 Interacting Protein 1 fragment, PSIP1 fragment protein, PC4 And SFRS1 Interacting Protein 1, LEDGF protein.

PSIP is involved in lens epithelial cell gene regulation and stress responses. It is a transcriptional coactivator involved in neuroepithelial stem cell differentiation and neurogenesis. PSIP may play a role in differentiation of lens epithelial to fiber cell terminal and also may act as a protector during stress-induced apoptosis.

### PSIP protein in Disease

A chromosomal aberration involving PSIP1 is associated with pediatric Acute Myeloid Leukemia (AML) with intermediate characteristics between M2-M3 French-American-British (FAB) subtypes.

Catalog No.	Amino Acids	Expression System	Purity
80-1384 (fragment)	349-455 with 6x His tag	Baculovirus	>90% pure
80-1381 (full)	1-530 with 6x His tag	Baculovirus	>90% pure



## SLA Protein

Also known as: Soluble Liver Antigen, Liver Pancreas Antigen

Soluble Liver Antigen is a UGA suppressor tRNA-associated protein. This family consists of several eukaryotic and archaeal proteins which are related to the Homo sapiens soluble liver antigen/liver pancreas antigen (SLA/LP autoantigen). The function of SLA is unknown, however, it has been suggested that the protein may function as a serine hydroxymethyl-transferase and may be an important enzyme in the thus far poorly understood selenocysteine pathway.

### SLA protein in Disease

Auto-antibodies to soluble liver antigen (SLA) and to liver and pancreas antigen (LP) have been described as disease specific, occurring in about 30% of all patients with Autoimmune Hepatitis [PMID: 10801173].

Catalog No.	Amino Acids	Expression System	Purity
80-1380	1-501 with 6x His tag and GST tag	E.coli	>90% pure

## SP100 Protein

Also known as: Nuclear autoantigen Sp100, SP100 Nuclear Antigen, Nuclear dot-associated SP100 protein, Speckled 100 kDa protein

The SP100 and promyelocytic leukemia proteins (PML) are constituents of nuclear domains, known as nuclear dots (NDs), also known as nuclear bodies (NBs) or PML bodies. They are covalently modified by SUMO1, small ubiquitin-related protein.

### SP100 protein in Disease

Nuclear bodies play a role in autoimmunity, virus infections, and in the etiology of Acute Promyelocytic Leukemia.

Catalog No.	Amino Acids	Expression System	Purity
80-1387	1-474 with 6x His tag	E.coli	>90% pure

## SSB Protein

Also known as: Sjogren Syndrome Antigen B, SS-B protein, La protein, Sjogren Syndrome B protein, Autoantigen La

La is involved in diverse aspects of RNA metabolism, including binding and protecting 3-prime UUU (OH) elements of newly RNA polymerase III-transcribed RNA, processing 5-prime and 3-prime ends of pre-tRNA precursors, acting as an RNA chaperone, and binding viral RNAs associated with hepatitis C virus.

### SSB protein in Disease

La protein was originally defined by its reactivity with auto-antibodies from patients with Sjögren's syndrome and Systemic Lupus Erythematosus (SLE).

Catalog No.	Amino Acids	Expression System	Purity
80-1376	1-148 with GST tag	Baculovirus	>85% pure

## TG3 Protein

Also known as: Transglutaminase 3 protein, TG3 protein, TG(E) protein, TGase-3 protein, Protein-glutamine-gamma-glutamyltransferase protein

Transglutaminases are enzymes that catalyze the crosslinking of proteins by epsilon-gamma glutamyl lysine isopeptide bonds. Transglutaminase 3 (TG3), also known as epidermal Transglutaminase (Tgase E) is involved in the formation of the cornified envelope in skin keratinocytes. It functions to cross-link structural proteins during epidermal terminal differentiation. TG3 activation requires proteolysis of the 77 kDa zymogen into two fragments of approximately 50 and 27 kDa to form the active enzyme.

### TG3 protein in Disease

Mutations in TG3 cause malformation of skin keratinocytes and has been implicated as the dominant autoantigen in Dermatitis Herpetiformis.

Catalog No.	Amino Acids	Expression System	Purity
80-1386	1-693 with 6x His tag	Baculovirus	>90% pure

## TPO Protein

Also known as: Thyroid Peroxidase protein, TDH2A protein, TPX protein, Thyroid Microsomal Antigen

Thyroid peroxidase or thyroperoxidase (TPO) acts as an enzyme and plays a central role in thyroid gland function. It is expressed mainly in the thyroid that liberates iodine for addition onto tyrosine residues on thyroglobulin for the production of the thyroid hormones, thyroxine (T<sub>4</sub>) or triiodothyronine (T<sub>3</sub>). TPO is stimulated by TSH, which upregulates gene expression.

### TPO protein in Disease

Thyroid peroxidase is a frequent epitope of anti-thyroid peroxidase antibodies (anti-TPO antibodies), auto-antibodies in autoimmune thyroid disease. This is most commonly associated with Hashimoto's Thyroiditis.

Catalog No.	Amino Acids	Expression System	Purity
80-1382	16-837 with 6x His tag	Baculovirus	>85% pure

## TRIM21 Protein

Also known as: 52 KDa Ribonucleoprotein Autoantigen Ro, Ro-52 protein, TRIM21 protein, RNF81 protein, R052 protein, SSA protein, SSA1 protein, E3 ubiquitin-protein ligase TRIM2, Tripartite motif-containing protein 21

TRIM21 is an intracellular antibody effector in the intracellular antibody-mediated proteolysis pathway. It binds to immunoglobulin G as well as immunoglobulin M on antibody marked non-enveloped virions which have infected the cell. It is responsible for directing the virions to the proteasome by autoubiquitination or by ubiquitination of a cofactor. Both the viral capsid and the bound antibody are degraded by the proteasome while TRIM21 remains intact.

### TRIM21 protein in Disease

TRIM21 is part of the RoSSA ribonucleoprotein, which includes a single polypeptide and one of four small RNA molecules. The RoSSA particle localizes to both the cytoplasm and the nucleus. RoSSA interacts with autoantigens in patients with Sjögren's syndrome and Systemic Lupus Erythematosus (SLE).

Catalog No.	Amino Acids	Expression System	Purity
80-1385	1-285 with 6x His tag	E.coli	>90% pure

## TROVE2 Protein

Also known as: 60 KDa Ribonucleoprotein R0, Ro60 protein, Trove-2 protein, TROVE Domain Family Member 2 protein, SSA2 protein

TROVE2 (TROVE domain family, member 2) is a protein-coding gene, and is affiliated with the lncRNA class. It functions as an RNA-binding protein that binds to misfolded non-coding RNAs, pre-5S rRNA, and several small cytoplasmic RNA molecules known as Y RNAs. It may also be involved in stabilization of some of these RNAs and protect them from degradation.

### TROVE2 protein in Disease

Diseases associated with TROVE2 include Sjögren's syndrome and Sialadenitis. It has also been implicated in Alcoholism via a related super-pathway.

Catalog No.	Amino Acids	Expression System	Purity
80-1383	1-538 with 6x His tag and GST tag	E.coli	>80% pure

## tTG Protein

Also known as: Tissue transglutaminase protein, TGM2 protein, G-ALPHA-h protein, GNAH protein, TG2 protein, TG protein

tTG or tissue transglutaminase is a 78-kDa, calcium dependent enzyme of the protein-glutamine  $\gamma$ -glutamyltransferases family. It is thought to be involved in the regulation of the cytoskeleton by crosslinking various cytoskeletal proteins including myosin, actin, and spectrin. Like other transglutaminases, it crosslinks proteins between an  $\epsilon$ -amino group of a lysine residue and a  $\gamma$ -carboxamide group of glutamine residue, creating an inter- or intramolecular bond that is highly resistant to proteolysis (protein degradation). It also functions in deamidation, GTP-binding / hydrolyzing, and isopeptidase activities.

### tTG protein in Disease

tTG is particularly known for being the autoantigen in Celiac disease (Coeliac disease), an illness in which the consumption of dietary gluten causes a pathological immune response resulting in the inflammation of the small intestine and subsequent villous atrophy.

Catalog No.	Amino Acids	Expression System	Purity
80-1379	1-687 with 6x His tag	E.coli	>85% pure



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