

**I. ORIGINAL ARTICLES**

1. Tryggvason, K. and Kouvalainen, K.: Numbers of nephrons in normal human kidneys and kidneys of patients with the congenital nephrotic syndrome. *Nephron* 15, 62-68, 1975.
2. Tryggvason, K.: Composition of the glomerular basement membrane in the congenital nephrotic syndrome of the Finnish type. *Eur. J. Clin. Invest.* 7, 177-180, 1977.
3. Risteli, J., Tryggvason, K. and Kivirikko, K.I.: Prolyl 3-hydroxylase: Partial characterization of the enzyme from rat kidney cortex. *Eur. J. Biochem.* 73, 485-492, 1977.
4. Tryggvason, K., Risteli, J. and Kivirikko, K.I.: Separation of prolyl 3-hydroxylase and 4-hydroxylase activities and 4-hydroxyproline requirement for synthesis of 3-hydroxyproline. *Biochem. Biophys. Res. Commun.* 76, 275-281, 1977.
5. Tryggvason, K., Risteli, J. and Kivirikko, K.I.: Glomerular basement membrane collagen and activities of the intracellular enzymes of collagen biosynthesis in congenital nephrotic syndrome of the Finnish type. *Clin. Chem. Acta* 82, 233-240, 1978.
6. Risteli, J., Tryggvason, K. and Kivirikko, K.I.: A rapid assay for prolyl 3-hydroxylase activity. *Anal. Biochem.* 84, 423-431, 1978.
7. Tryggvason, K. and Kivirikko, K.I.: Heterogeneity of pepsin-solubilized human basement membrane collagen. *Nephron* 21, 230-235, 1978.
8. Risteli, J., Tuderman, L., Tryggvason, K. and Kivirikko, K.I.: Effect of hepatic injury on prolyl 3-hydroxylase and 4-hydroxylase concentration in liver and serum. *Biochem. J.* 170, 129-135, 1978.
9. Tryggvason, K.: Morphometric studies on glomeruli in the congenital nephrotic syndrome. *Nephron* 22, 544-550, 1978.
10. Tryggvason, K., Majamaa, K. and Kivirikko, K.I.: Prolyl 3-hydroxylase and 4-hydroxylase activities in the rat. *Biochem. J.* 178, 127-131, 1979.
11. Tryggvason, K., Gehron-Robey, P. and Martin, G.R.: Biosynthesis of type IV procollagens. *Biochemistry* 19, 1284-1289, 1979.
12. Tryggvason, K., Majamaa, K., Risteli, J. and Kivirikko, K.I.: Partial purification and characterization of chick-embryo prolyl 3-hydroxylase. *Biochem. J.* 183, 303-307, 1980.
13. Liotta, L.A., Tryggvason, K., Garbisa, S., Foltz, C.M. and Shafie, S.: Metastatic potential correlates with enzymatic degradation of basement membrane collagen. *Nature* 284, 67-68, 1980.
14. Garbisa, S., Kniska, K., Tryggvason, K., Foltz, C. and Liotta, L.A.: Quantitation of basement membrane collagen degradation by living tumor cells in vitro. *Cancer Lett.* 9, 359-366, 1980.
15. Garbisa, S., Tryggvason, K., Foidart, J.M. and Liotta, L.A.: Assay for radioactive type IV collagen in the presence of other proteins using a specific collagenase. *Anal. Biochem.* 107, 187-192, 1980.
16. Foidart, J.M., Tryggvason, K., Gehron-Robey, P., Liotta, L.A. and Martin, G.R.: Biosynthesis of type IV and V collagens by human placenta. *Coll. Res.* 1, 137-150, 1981.
17. Liotta, L.A., Tryggvason, K., Garbisa, S., Gehron-Robey, P. and Abe, S.: Partial purification and characterization of a neutral protease which cleaves type IV collagen. *Biochemistry* 20, 100-104, 1981.
18. Myllylä, R., Tryggvason, K., Kivirikko, K.I. and Reddi, A.H.: Changes in intracellular enzymes of collagen biosynthesis during matrix-induced cartilage and bone development. *Biochem. Biophys. Acta* 674, 233-239, 1981.
19. Tryggvason, K., Oikarinen, J., Viinikka, L. and Ylikorkala, O.: Effects of laminin, proteoglycan and type IV collagen, components of basement membranes, on platelet aggregation. *Biochem. Biophys. Res. Commun.* 100, 233-239, 1981.

20. Garbisa, S., Liotta, L.A., Tryggvason, K. and Siegel, G.P.: Antibodies to collagenase-resistant terminal regions on type IV procollagen recognize whole basement membrane and 7-S collagen. *FEBS Lett.* 127, 257-262, 1981.
21. Oikarinen, A., Savolainen, E-R., Tryggvason, K., Foidart, J.M. and Kiistala, U.: Basement membrane components and galactosylhydroxylsyl glycosyltransferase in suction blisters of human skin. *Br. J. Derm.* 106, 257-266, 1982.
22. Pihlajaniemi, T., Myllylä, R., Kivirikko, K.I. and Tryggvason, K.: Effects of streptozotocin diabetes, glucose and insulin on the metabolism of type IV collagen and proteoglycan in murine basement membrane-forming tissue. *J. Biol. Chem.* 257, 14914-14920, 1982.
23. Salo, T., Liotta, L.A., Keski-Oja, J., Turpeenniemi-Hujanen, T. and Tryggvason, K.: Secretion of basement membrane collagen degrading enzyme and plasminogen activator by transformed cells - Role in metastasis. *Int. J. Cancer* 30, 669-673, 1982.
24. Kleinman, H.K., McGarvey, M.L., Liotta, L.A., Gehron-Robey, P., Tryggvason, K. and Martin, G.R.: Isolation and characterization of native type IV collagen, laminin and heparan sulfate proteoglycan from EHS sarcoma. *Biochemistry* 21, 6188-6193, 1982.
25. Salo, T., Liotta, L.A. and Tryggvason, K.: Purification and characterization of a murine basement membrane collagen degrading enzyme secreted by metastatic tumor cells. *J. Biol. Chem.* 258, 3058-3063, 1983.
26. Palotie, A., Tryggvason, K., Peltonen, L. and Seppä, H.: Components of subendothelial aorta basement membrane: Immunohistochemical localization and role in cell adhesion. *Lab. Invest.* 49, 362-370, 1983.
27. Isotalo, H., Tryggvason, K., Vierikko, P. and Vihko, R.: Plasminogen activator activities and estrogen and progesterin receptor levels in normal human tissues and malignant tumors of breast and ovarium. *Anticancer Res.* 3, 331-336, 1983.
28. Höyhtyä, M., Myllylä, R., Piuva, J., Kivirikko, K.I. and Tryggvason, K.: Monoclonal antibodies to human prolyl 4-hydroxylase. *Eur. J. Biochem.* 141, 477-482, 1984.
29. Fessler, L., Duncan, K., Fessler, J.H., Salo, T. and Tryggvason, K.: Characterization of the procollagen IV cleavage products produced by a specific tumor collagenase. *J. Biol. Chem.* 259, 9783-9789, 1984.
30. Pihlajaniemi, T., Tryggvason, K., Myers, J., Kurkinen, M., Lebo, R., Cheung, M., Prockop, D.J. and Boyd, C.D.: cDNA clones for the pro- $\alpha$ 1(IV) chain of human type IV procollagen reveal an unusual homology of amino acid sequences in two halves of the carboxyl-terminal domain. *J. Biol. Chem.* 260, 7681-7687, 1985.
31. Salo, T., Turpeenniemi-Hujanen, T. and Tryggvason, K.: Tumor-promoting phorbol esters and cell proliferation stimulate secretion of basement membrane (type IV) collagen-degrading metalloproteinase by human fibroblasts. *J. Biol. Chem.* 260, 8526-8531, 1985.
32. Soininen, R., Tikka, L., Chow, L., Pihlajaniemi, T., Kurkinen, M., Prockop, D.J., Boyd, C.D. and Tryggvason, K.: Large introns in the 3'-end of the gene for the pro $\alpha$ 1(IV) chain of human basement membrane collagen. *Proc. Natl. Acad. Sci. USA* 83, 1568-1572, 1986.
33. Soininen, R., Chow, L., Kurkinen, M., Tryggvason, K., and Prockop, D.J.: The gene for the  $\alpha$ 1(IV) chain of human type IV procollagen: the exon structures do not coincide with the two structural subdomains in the globular carboxy-terminus of the protein. *EMBO J.*, 5, 2821-2823, 1986
34. Boyd, C.D., Weliky, K., Deak, S.B., Christiano, A.M., Mackenzie, J.M., Sandell, L., Tryggvason, K. and Magenis, E.: The single copy gene coding for human  $\alpha$ 1(IV) procollagen is located at the terminal end of the long arm of chromosome 13. *Human Gen.* 74, 121-125, 1986.

35. Höyhtyä, M., Vihko, P., Vuolas, L., Tryggvason, K. and Vihko, R.: High-affinity monoclonal antibodies specific for human prostatic acid phosphatase. *Clin. Chem.* 33, 103-107, 1987.
36. Oikarinen, A., Salo, T. and Tryggvason, K.: Dexamethasone modulates the metabolism of type IV collagen and fibronectin in human basement membrane forming fibrosarcoma (HT-1080) cells. *Biochem. J.* 245, 235-241, 1987.
37. Hostikka, S.L., Kurkinen, M. and Tryggvason, K.: Nucleotide sequence coding for the human type IV collagen  $\alpha 2$  chain cDNA reveals extensive homology with the NC-1 domain of  $\alpha 1(\text{IV})$ , but not with the collagenous domain or untranslated region. *FEBS Lett.* 216, 281-286, 1987.
38. Tikka, L., Roiko, K., Soininen, R., Prockop, D.J. and Tryggvason, K.: A Hind III polymorphism in the 3' end of the human  $\alpha 1(\text{IV})$  collagen gene. *Nucl. Acids. Res.* 15, 5497, 1987.
39. Pikkarainen, T., Eddy, R., Fukushima, Y., Byers, M., Shows, T., Pihlajaniemi, T., Saraste, M. and Tryggvason, K.: Human laminin B1 chain: a multidomain protein with gene (LAMB1) locus in the q22 region of chromosome 7. *J. Biol. Chem.* 262, 10454-10462, 1987.
40. Hostikka, S.L. and Tryggvason, K.: Extensive structural differences between genes for the  $\alpha 1$  and  $\alpha 2$  chains of type IV collagen despite conservation of coding sequences. *FEBS Lett.* 224, 297-305, 1987.
41. Soininen, R., Haka-Risku, T., Prockop, D.J. and Tryggvason, K.: Complete primary structure of the  $\alpha 1$  chain of human basement membrane (type IV) collagen. *FEBS Lett.* 225, 188-194, 1987.
42. Pikkarainen, T., Kallunki, T. and Tryggvason, K.: Human laminin B2 chain: Comparison of the complete amino acid sequence with the B1 chain reveals variability in sequence homology between different structural domains. *J. Biol. Chem.* 263, 6751-6758, 1988.
43. Pajunen, L., Myllylä, R., Helaakoski, T., Pihlajaniemi, T., Tasanen, K., Höyhtyä, M., Tryggvason, K., Solomon, E. and Kivirikko, K.I.: Assignment of the gene coding for both the  $\beta$ -subunit of prolyl 4-hydroxylase and the enzyme disulfide isomerase to human chromosome region 17p11 -> qter. *Cytogen. Cell Genet.* 47, 37-41, 1988.
44. Höyhtyä, M., Turpeenniemi-Hujanen, T., Stetler-Stevenson, W., Krutzsch, H., Tryggvason, K. and Liotta, L.A.: Monoclonal antibodies to type IV collagenase recognize a protein with limited sequence homology to interstitial collagenase and stromelysin. *FEBS Lett.* 233, 109-113, 1988.
45. Tikka, L., Pihlajaniemi, T., Henttu, P., Prockop, D.J. and Tryggvason, K.: Gene structure for the  $\alpha 1$  chain of human short chain collagen (type XIII) with alternatively spliced transcripts and the translation termination codon at the 5' end of the last exon. *Proc. Natl. Acad. Sci. USA* 85, 7491-7495, 1988.
46. Fukushima, Y., Pikkarainen, T., Kallunki, T., Eddy, R.L., Byers, M.G., Hayley, L.L., Henry, W.M., Tryggvason, K. and Shows, T.B.: Isolation of human laminin B2 (LAMB2) cDNA clone and assignment of the gene to the chromosome 1q25-q31 region. *Cytogen. Cell Genet.* 48, 137-141, 1988.
47. Soininen, R., Huotari, M., Hostikka, S.L., Prockop, D.J. and Tryggvason, K.: The structural genes for  $\alpha 1$  and  $\alpha 2$  chains of human type IV collagen are divergently encoded on opposite DNA strands and have an overlapping promoter region. *J. Biol. Chem.* 263, 17217-17220, 1988.
48. Hostikka, S.L. and Tryggvason, K.: The complete primary structure of the  $\alpha 2$  chain of human type IV collagen and comparison with the 1(IV) chain. *J. Chem Biol.* 263, 19488-19493, 1988.
49. Ikonen, J., Pikkarainen, T., Savolainen, E.R. and Tryggvason, K.: A Hpa I polymorphism in the human laminin B1 chain gene on 7q22. *Nucleic. Acids. Res.* 17, 473, 1989.
50. Soininen, R., Huotari, M., Ganguly, A., Prockop, D.J. and Tryggvason, K.: Structural organization of the gene for the  $\alpha 1(\text{IV})$  chain of human type IV collagen. *J. Biol. Chem.* 264, 13565-13571, 1989.

51. Kallunki, T., Pikkarainen, T., Tryggvason, K. and Savolainen, E.-R.: A Pst I polymorphism in the human laminin B2 chain gene on 1q25-q3. *Nucleic Acids Res.* 17, 4423, 1989.
52. Pikkarainen, T., Savolainen, E.-R. and Tryggvason, K.: Nhe I and Hinc II polymorphisms in the human laminin B1 chain gene on 7q22. *Nucleic Acids Res.* 17, 4424, 1989.
53. Murphy, G., Ward, R., Hembry, R.M., Reynolds, J.J., Kühn, K. and Tryggvason, K.: Characterization of gelatinase from pig polymorphonuclear leucocytes. A metalloproteinase resembling tumour type IV collagenase. *Biochem. J.* 258, 463-472, 1989.
54. Shows, T., Tikka, I., Byers, M.G., Eddy, R.L., Haley, L.L., Henry, W.M., Prockop, D.J. and Tryggvason, K.: Assignment of the human collagen  $\alpha 1$ (XIII) chain gene (*COL13A1*) to the q22 region of chromosome 10. *Genomics* 5, 128-133, 1989.
55. Huhtala, P., Eddy, R.L., Fan, Y.S., Byers, M.G., Shows, T. B. and Tryggvason, K.: Completion of the primary structure of the human type IV collagenase preproenzyme and assignment of the gene (*CLG4*) to the q21 region of chromosome 16. *Genomics* 6, 554-559, 1990.
56. Hostikka, S.L., Eddy, R.L., Byers, M.G., Höyhtyä, M., Shows, T.B. and Tryggvason, K.: Identification of a distinct type IV collagen  $\alpha$  chain with restricted kidney distribution and assignment of the gene to the locus of X chromosome-linked Alport syndrome. *Proc. Natl. Acad. Sci. USA* 87, 1606-1610, 1990.
57. Huhtala, P., Chow, L. and Tryggvason, K.: Structure of the human type IV collagenase gene. *J. Biol. Chem.* 265, 11077-11082, 1990.
58. Höyhtyä, M., Hujanen, E., Turpeenniemi-Hujanen, T., Thorgeirsson, U., Liotta, L.A. and Tryggvason, K.: Modulation of type-IV collagenase activity and invasive behavior of metastatic human melanoma (A-2058) cells *in vitro* by monoclonal antibodies to type IV-collagenase. *Int. J. Cancer* 46, 282-286, 1990.
59. Barker, D., Hostikka, S.L., Zhou, J., Chow, L.T., Oliphant, A.R., Gerken, S.C., Gregory, M.C., Skolnick, M.H., Atkin, C.L. and Tryggvason, K.: Identification of mutations in the COL4A5 collagen gene in Alport syndrome. *Science* 248, 1224-1227, 1990.
60. Vuolteenaho, R., Chow, L. and Tryggvason, K.: Structure of the human laminin B1 chain gene. *J. Biol. Chem.* 265, 15611-15616, 1990.
61. Kallunki, T., Ikonen, J., Chow, L., Kallunki, P. and Tryggvason, K.: Structure of the human laminin B2 chain gene reveals extensive divergence from the laminin B1 chain gene. *J. Biol. Chem.* 266, 221-228, 1991.
62. Zhou, J., Hostikka, S.L., Chow, L.T. and Tryggvason, K.: Characterization of the 3' half of the human type IV collagen  $\alpha 5$  gene that is affected in Alport syndrome. *Genomics* 9, 1-9, 1991.
63. Zhou, J., Barker, D., Hostikka, S.L., Gregory, M., Atkin, C. and Tryggvason, K.: Single base mutation in  $\alpha 5$ (IV) collagen chain gene converting a conserved cysteine to serine in Alport syndrome. *Genomics* 9, 10-18, 1991.
64. Nissinen, M., Vuolteenaho, R., Boot-Handford R., Kallunki, P. and Tryggvason, K.: Primary structure of the human laminin A chain: Limited expression in human tissues. *Biochem. J.* 276, 369-379, 1991.
65. Huhtala, P., Tuuttila, A., Chow, L., Lohi, J., Keski-Oja, J. and Tryggvason, K.: Complete structure of the human gene for 92-kDa type IV collagenase: Divergent effects of TPA and TGF- $\beta 1$  on transcription of the 92-kDa and 70-kDa enzyme genes in HT-1080 cells. *J. Biol. Chem.* 266, 16485-16490, 1991.
66. Kallunki, P., Eddy, R.L., Byers, M.G., Kestilä, M., Shows, T.B. and Tryggvason, K.: Cloning of human heparan sulfate proteoglycan core protein, assignment of the gene (*HSPG1*) to 1p36.1->p35 and identification of a Bam HI restriction fragment length polymorphism. *Genomics* 11, 389-396, 1991.

67. Tikka, L., Elomaa, O., Pihlajaniemi, T. and Tryggvason, K.: Human  $\alpha 1$  (XIII) collagen gene: multiple forms of the gene transcripts are generated through complex alternative splicing of several short exons. *J. Biol. Chem.* 266, 17713-17719, 1991.
68. Barker, D.F., Fain, P.R., Goldgar, D.E., Dietz-Band, J.N., Turco, A.E., Kashtan, C.E., Gregory, M.C., Tryggvason, K., Skolnick, M.H. and Atkin, C.L.: High density genetic and physical mapping of DNA markers near the X-linked Alport syndrome locus: Definition and use of flanking polymorphic markers. *Human Genet.* 88, 189-194, 1991.
69. Pyke, C., Ralfkiær, E., Huhtala, P., Hurskainen, T., Danø, K. and Tryggvason, K.: Localization of messenger RNA for Mr 72,000 and 92,000 type IV collagenases in human skin cancers by *in situ* hybridization. *Cancer Res.* 52, 1336-1341, 1992.
70. Kallunki, P. and Tryggvason, K.: Human basement membrane heparan sulfate proteoglycan core protein: A 467-kD protein containing multiple domains resembling elements of the low density lipoprotein receptor, laminin, neural cell adhesion molecules, and epidermal growth factor. *J. Cell. Biol.* 116, 559-571, 1992.
71. Reponen, P., Sahlberg, C., Huhtala, P., Hurskainen, T., Thesleff, I. and Tryggvason, K.: Molecular cloning of murine 72-kDa type IV collagenase and its expression during mouse development. *J. Biol. Chem.* 267, 7856-7862, 1992.
72. Autio-Harmainen, H., Hurskainen, T., Niskasaari, K., Höyhty, M. and Tryggvason, K.: Simultaneous expression of 70 kilodalton type IV collagenase and type IV collagen  $\alpha 1$ (IV) chain genes by cells of early human placenta and gestational endometrium. *Lab. Invest.* 67, 191-200, 1992.
73. Zhou, J., Hertz, J.M. and Tryggvason, K.: Mutation in the  $\alpha 5$ (IV) collagen chain in juvenile-onset Alport syndrome without hearing loss or ocular lesions: Detection by denaturing gradient gel electrophoresis of a PCR product. *Am. J. Human Genet.* 50, 1291-1300, 1992.
74. Zhou, J., Herz, J.M., Leinonen, A. and Tryggvason, K.: Complete amino acid sequence of the human  $\alpha 5$ (IV) collagen chain and identification of a single base mutation in exon 23 converting glycine-521 in the collagenous domain to cysteine in an Alport syndrome patient. *J. Biol. Chem.* 267, 12475-12481, 1992.
75. Keski-Oja, J., Lohi, J., Tuuttila, A., Tryggvason, K. and Vartio, T.: Proteolytic processing of the 72,000-Da type IV collagenase by urokinase plasminogen activator. *Exp. Cell Res.* 202, 471-476, 1992.
76. Pikkarainen, T., Schulthess, T., Engel, J. and Tryggvason, K.: Recombinant laminin B1 chains exhibit intact short-arm domains but do not form oligomeric molecules. *Eur. J. Biochem.* 209, 571-582, 1992.
77. Kallunki, P., Sainio, K., Eddy, R., Byers, M., Kallunki, T., Sariola, H., Beck, K., Hirvonen, H., Shows, T.B. and Tryggvason, K.: A truncated laminin chain homologous to the B2 chain: structure, spatial expression and chromosomal assignment. *J. Cell Biol.* 119, 679-693, 1992.
78. Smeets, H.J.M., Melenhorst, J.J., Lemmink, H.H., Schröder, C.H., Nelen, M.R., Zhou, J., Hostikka, S.L., Tryggvason, K., Ropers, H.-H., Jansweijer, M.C.E., Monnens, L.A.H., Brunner, H.G., van Oost, B.A.: Different mutations in the COL4A5 collagen gene in two patients with different features of Alport Syndrome. *Kidney Int.* 42, 83-88, 1992.
79. Knebelmann, B., Deschenes, G., Gros, F., Hors, M.-C., Grünfeld, J.-P., Zhou, J., Tryggvason, K., Gubler, M.-C. and Antignac, C.: Substitution of arginine for glycine 325 in the collagen  $\alpha 5$ (IV) chain associated with X-linked Alport syndrome: Characterization of the mutation by direct sequencing of PCR-amplified lymphoblast cDNA fragments. *Am. J. Hum. Genet.* 51, 135-142, 1992.

80. Netzer, K.-O., Renders, L., Zhou, J., Pullig, O., Tryggvason, K. and Weber, M.: Deletions of the *COL4A5*-gene in patients with Alport syndrome. *Kidney Int.* 42, 1336-1344, 1992.
81. M'Rad, R., Sanak, M., Deschenes, G., Zhou, J., Bonaiti-Pellie, C., Holvoet-Vermaut, L., Heuertz, S., Gubler, M.-C., Broyer, M., Grunfeld, J.-P., Tryggvason, K. and Hors-Cayla, M.-C.: Alport syndrome: a genetic study of 31 families. *Hum. Genet.* 90, 420-426, 1992.
82. Antignac, C., Zhou, J., Sanak, M., Cochat, P., Roussel, B., Deschenes, G., Gros, F., Knebelmann, B., Hors-Cayla, M.C., Tryggvason, K. and Gubler, M.C.: Alport syndrome and diffuse leiomyomatosis - Deletions in the 5' end of the *COL4A5* collagen gene. *Kidney Int.* 42, 1178-1183, 1992.
83. Sahlberg, C., Reponen, P., Tryggvason, K. and Thesleff, I.: Association between the expression of murine 72 kDa type IV collagenase by odontoblasts and basement membrane degradation during mouse tooth development. *Arch. Oral Biol.* 37, 1021-1030, 1992.
84. Tryggvason, K., Zhou, J., Hostikka, S.L. and Shows, T.: Molecular genetics of Alport syndrome. *Kidney Int.* 43, 38-44, 1992.
85. Kleiner, D.E., Tuuttila, A., Tryggvason, K. and Stetler-Stevenson, W.G.: Stability analysis of latent and active 72-kDa type IV collagenase: The role of tissue inhibitor of metalloproteinases-2 (TIMP-2). *Biochemistry* 32, 1583-1592, 1993.
86. Netzer, K.-O., Pullig, O., Zhou, J., Frei, U., Tryggvason, K. and Weber, M.: *COL4A5* splice site mutation and  $\alpha 5(\text{IV})$  collagen mRNA in Alport syndrome. *Kidney Int.* 43, 486-492, 1993.
87. Pyke, C., Ralfkiaer, E., Tryggvason, K. and Danø, K.: Messenger RNA for two type IV collagenases is located in stromal cells in human colon cancer. *Am. J. Pathol.* 142, 359-365, 1993.
88. Zhou, J., Gregory, M.C., Hertz, J.M., Barker, D.F., Atkin, C., Spencer, E. and Tryggvason, K.: Mutations in the codon for a conserved arginine-1563 in the *COL4A5* collagen gene in Alport syndrome. *Kidney Int.* 43, 722-729, 1993.
89. Lemmink, H.H., Schröder, C.H., Brunner, H.G., Nelen, M.R., Zhou, J., Tryggvason, K., Haagsma-Schouten, W.A.G., Roodvoets, A.P., Rascher, W., van Oost, B.A. and Smeets, J.M.: Systematic screening for mutations in 16 exons of the *COL4A5* gene in Alport syndrome. *Genomics* 17, 485-489, 1993.
90. Zhou, J., Mochizuki, T., Smeets, H., Antignac, C., Laurila, P., de Paepe, A., Tryggvason, K. and Reeders, S.T.: Deletion of the paired  $\alpha 5(\text{IV})$  and  $\alpha 6(\text{IV})$  collagen genes in inherited smooth muscle tumors. *Science* 261, 1167-1169, 1993.
91. Autio-Harmainen, H., Karttunen, T., Hurskainen, T., Höyhty, M., Kauppila, A. and Tryggvason, K.: Expression of 72 kilodalton type IV collagenase (gelatinase A) in benign and malignant ovarian tumors. *Lab. Invest.* 69, 312-321, 1993.
92. Heikkilä, P., Soininen, R. and Tryggvason, K.: Directional regulatory activity of *cis*-acting elements in the bidirectional  $\alpha 1(\text{IV})$  and  $\alpha 2(\text{IV})$  collagen gene promoter. *J. Biol. Chem.* 268, 24677-24682, 1993.
93. Vuolteenaho, R., Nissinen, M., Saino, K., Byers, M., Eddy, R., Hirvonen, H., Shows, T.B., Sariola, H., Engvall, E. and Tryggvason, K.: Human laminin M chain (merosin): Complete primary structure, chromosomal assignment and expression of the M and A chain in human fetal tissues. *J. Cell Biol.* 124, 381-394, 1994.
94. Kestilä, M., Männikkö, M., Holmberg, C., Tryggvason, K. and Peltonen, L.: Congenital nephrotic syndrome of the Finnish type is not associated with the *Pax-2* gene despite the promising transgenic animal model. *Genomics* 19, 570-572, 1994.
95. Pulkkinen, L., Christiano, A.M., Airene, T., Haakana, H., Tryggvason, K. and Uitto, J.: Mutations in the  $\gamma 2$  chain gene (*LAMC2*) of kalinin/laminin 5 in the junctional forms of epidermolysis bullosa. *Nature Genetics* 6, 293-298, 1994.

96. Aberdam, D., Galliano, M.-F., Vailly, J., Pulkkinen, L., Bonifas, J., Christiano, A.M., Tryggvason, K., Uitto, J., Epstein, E.H. Jr., Ortonne, J.-P. and Meneguzzi, G.: Herlitz's junctional epidermolysis bullosa is linked to mutations in the gene (*LAMC2*) for the  $\gamma 2$  subunit of nicein/kalinin (Laminin-5). *Nature Genetics* 6, 299-304, 1994.
97. Zhou, J., Leinonen, A. and Tryggvason, K.: Structure of the human type IV collagen *COL4A5* gene. *J. Biol. Chem.* 269, 6608-6614, 1994.
98. Reponen, P., Sahlberg, C., Munaut, C., Thesleff, I. and Tryggvason, K.: High expression of 92-kDa type IV collagenase (gelatinase B) in the osteoclasts lineage during mouse development. *J. Cell. Biol.* 124, 1091-1102, 1994.
99. Kestilä, M., Männikkö, M., Holmberg, C., Korpela, K., Savolainen, E.-R., Peltonen, L. and Tryggvason, K.: Exclusion of eight genes coding for major basement membrane components as mutated loci in congenital nephrotic syndrome of the Finnish type. *Kidney Int.* 45, 986-990, 1994.
100. Kestilä, M., Männikkö, M., Holmberg, C., Gyapay, G., Weissenbach, J., Savolainen, E.-R., Peltonen, L. and Tryggvason, K.: Congenital nephrotic syndrome of the Finnish type maps to the long arm of chromosome 19. *Am. J. Hum. Genet.* 54, 757-764, 1994.
101. Antignac, C., Knebelmann, B., Drouot, L., Deschenes, G., Hors-Cayla, M.-C., Zhou, J., Tryggvason, K., Grünfeld, J.-P., Broyer, M. and Gubler, M.-C.: Deletions in the *COL4A5* collagen gene in X-linked Alport syndrome: Characterization of the Pathological transcripts in non renal cells and correlation with disease expression. *J. Clin. Invest.* 93, 1195-1207, 1994.
102. Hujanen, E.S., Väisänen, A., Zheng, A., Tryggvason, K. and Turpeenniemi-Hujanen, T.: Modulation of Mr 72,000 and Mr 92,000 type-IV collagenases (gelatinases A and B) gene expression by interferon alpha and gamma in human melanoma. *Int. J. Cancer* 58, 582-586, 1994.
103. Mariyama, M., Leinonen, A., Mochizuki, T., Tryggvason, K. and Reeders, S.: Complete primary structure of the human  $\alpha 3(\text{IV})$  collagen chain: Coexpression of the  $\alpha 3(\text{IV})$  and  $\alpha 4(\text{IV})$  collagen chains in human tissues. *J. Biol. Chem.* 269, 23013-23017, 1994.
104. Pyke, C., Rømer, J., Kallunki, P., Lund, L.R., Ralfkiær, E., Danø, K. and Tryggvason, K.: Laminin  $\gamma 2$  chain is preferentially expressed in invading malignant cells in human cancers. *Am. J. Pathol.* 145, 782-791, 1994.
105. Leinonen, A., Mariyama, M., Mochizuki, T., Tryggvason, K. and Reeders, S.: Complete primary structure of the human type IV collagen  $\alpha 4(\text{IV})$  chain: Comparison with structure and expression of the other  $\alpha(\text{IV})$  chains. *J. Biol. Chem.* 269, 26172-26177, 1994.
106. Ding, J., Zhou, J., Tryggvason, K. and Kashtan, C.E.: *COL4A5* deletions in three patients with Alport syndrome and post-transplant anti-GBM nephritis. *J. Am. Soc. Nephrol.* 5, 161-168, 1994.
107. Carome, M.A., Striker, L.J., Peten, E.P., Elliot, S.J., Stetler-Stevenson, W.G., Reponen, P., Tryggvason, K. and Striker, G.E.: Assessment of 72 kDa gelatinase and tissue inhibitor of metalloproteinase-1 (TIMP-1) gene expression in normal and sclerotic murine glomeruli *in vivo*. *J. Am. Soc. Nephrol.* 5, 1391-1399, 1994.
108. Iivanainen, A., Vuolteenaho, R., Sainio, K., Janit-Sait, S., Eddy, R., Shows, T.B., Sariola, H. and Tryggvason, K.: The human laminin  $\beta 2$  chain (s-laminin): Structure, expression in fetal tissues and chromosomal assignment of the *LAMB2* gene. *Matrix Biol.* 14, 489-497, 1994.
109. Hertz, J.M., Heiskari, N., Zhou, J., Jensen, U.B. and Tryggvason, K.: A nonsense mutation in the *COL4A5* gene in a family with X-linked Alport syndrome: Carrier detection and prenatal diagnosis. *Kidney Int.* 47, 327-332, 1995.
110. Reponen, P., Leivo, I., Sahlberg, C., Apte, S.S., Olsen, B.R., Thesleff, I. and Tryggvason, K.: The 92-kDa type IV collagenase and TIMP-3, but the 72-kDa

- type IV collagenase, or TIMP-2 are highly expressed during mouse embryo implantation. *Devel. Dynamics* 202, 388-396, 1995.
111. Bergmann, U., Tuuttila, A., Stetler-Stevenson, W.G. and Tryggvason, K.: Autolytic activation of recombinant human 72 kDa type IV collagenase (MMP-2, gelatinase A). *Biochemistry* 34, 2819-2825, 1995.
  112. Heidet, L., Dahan, K., Zhou, J., Zhang, X., Cochat, P., Gould, J.D.M., Leppig, K.A., Proesmans, W., Guyot, C., Guillot, M., Roussel, B., Tryggvason, K., Grünfeld, J.-P., Gubler, M.-C. and Anticnac, C. Deletions of both  $\alpha 5(\text{IV})$  and  $\alpha 6(\text{IV})$  collagen genes in Alport syndrome and in smooth muscle tumors. *Hum. Mol. Genet.* 4, 99-108, 1995.
  113. Elomaa, O., Kangas, M., Sahlberg, C., Tuukkanen, J., Sormunen, R., Liakka, A., Thesleff, I., Kraal, G. and Tryggvason, K.: Cloning of a novel bacteria-binding receptor, structurally related to scavenger receptors and expressed in a subset of macrophages. *Cell* 80, 603-609, 1995.
  114. Boye, E., Flinter, F., Zhou, J., Tryggvason, K., Bobrow, M. and Harris, A.: Detection of 12 novel mutations in the collagenous domain of the *COL4A5* in Alport syndrome. *Hum. Mut.* 5, 197-204, 1995.
  115. Iivanainen, A., Sainio, K., Sariola, H. and Tryggvason, K.: Primary structure and expression of a novel human laminin  $\alpha 4$  chain. *FEBS Lett.* 365, 183-188, 1995.
  116. Vailly, J., Pulkkinen, L., Christiano, A.M., Tryggvason, K., Uitto, J., Ortonne, J.P. and Meneguzzi, G.: Identification of a homozygous exon-skipping mutation in the *LAMC2* gene in a patient with Herlitz's junctional epidermolysis bullosa. *J. Invest. Derm.* 104, 434-437, 1995.
  117. Helbling-Leclerc, A., Zhang, X., Topaloglu, H., Cruaud, C., Tesson, F., Weissenbach, J., Tome, F.M.S., Schwartz, K., Fardeau, M., Tryggvason, K. and Guicheney, P.: Mutations in the laminin  $\alpha 2$ -chain gene (*LAMA2*) cause merosin-deficient congenital muscular dystrophy. *Nature Genetics* 11, 216-218, 1995.
  118. Pyke, C., Salo, S., Ralfkiær, E., Rømer, J., Danø, K. and Tryggvason, K.: Laminin-5 is a marker of invading cancer cells in some human carcinomas and is coexpressed with the receptor for urokinase plasminogen activator in budding cancer cells in colon adenocarcinomas. *Cancer Res.* 55, 4132-4139, 1995.
  119. Männikkö, M., Kestilä, M., Holmberg, C., Norio, R., Ryyänen, M., Olsen, A., Peltonen, L. and Tryggvason, K.: Fine mapping and haplotype analysis of the locus for congenital nephrotic syndrome locus on chromosome 19q13.1. *Am. J. Hum. Genet.* 57 1377-1383, 1995.
  120. Heikkilä, P., Parpala, T., Lukkariinen, O., Weber, M. and Tryggvason, K.: Adenovirus-mediated gene transfer into kidney glomeruli using an *ex vivo* and *in vivo* kidney perfusion system – First steps towards gene therapy of Alport syndrome. *Gene Therapy* 3, 21-27, 1996.
  121. Airene, T., Haakana, H., Sainio, K., Kallunki, T., Kallunki, P., Sariola, H. and Tryggvason, K.: Structure of the human laminin  $\gamma$  chain gene (*LAMC2*) and alternative splicing with different tissue distribution of two transcripts. *Genomics* 32, 54-64, 1996.
  122. Renieri, A., Bruttini, M., Galli, L., Zanelli, P., Neri, T., Rossetti, S., Turco, A., Heiskari, N., Zhou, J., Gusmano, R., Massella, L., Banfi, G., Scolari, F., Sessa, A., Rizzoni, G., Tryggvason, K., Pignatti, P.F., Savi, M., Ballabio, A., and de Marchi, M.: X-linked Alport Syndrome: a SSCP-based mutation survey over all 51 exons of the *COL4A5*. *Am. J. Hum. Genet.* 58:1192-1204, 1996.
  123. Nissinen, M., Helbling-Leclerc, A., Zhang, X., Evangelista T., Topaloglu, H., Cruaud C., Weissenbach, J., Fardeau, M., Tomé, F.M.S., Schwartz, K., Tryggvason, K. and Guicheney, P.: Substitution of a conserved cysteine-996 in a cysteine-rich motif of the laminin  $\alpha 2$  chain in congenital muscular dystrophy with partial deficiency of the protein. *Am. J. Hum. Genet.* 58, 1177-1184, 1996.



124. Heiskari, N., Zhang, X., Leinonen, A., Barker, D., Gregory, M., Atkin, G., Netzer, K.-O., Weber, M., Reeders, S., Grönhagen-Riska, C., Neumann, H.P.H., Trembath, R. and Tryggvason, K.: Identification of 17 mutations in ten exons in the *COL4A5* collagen gene, but no mutations found in four exons in *COL4A6*. A study of 250 patients with hematuria and suspected of having Alport syndrome. *J. Am. Soc. Nephrol.* 7, 702-709, 1996.
125. Zhang, X., Zhou, J., Reeders, S. T. and Tryggvason, K.: Structure of the human type IV collagen *COL4A6* gene which is mutated in Alport syndrome-associated leiomyomatosis. *Genomics* 33, 473-479, 1996.
126. Dehan, P., van der Heuvel, L.P., Smeets, H.J., Tryggvason, K., and Foidart, J.-M.: Identification of posttransplant anti- $\alpha 5(\text{IV})$  collagen alloantibodies in X-linked Alport syndrome. *Nephrol. Dial. Transpl.* 11, 1983-1988, 1996.
127. Dehan, P., Weber, M., Zhang, X., Reeders, S.T., Foidart, J.-M. and Tryggvason, K.: Sera from patients with anti-GBM nephritis including Goodpasture syndrome show heterogenous reactivity with recombinant NC1 domain of type IV collagen  $\alpha$  chains. *Nephrol. Dial. Transpl.* 11, 2215-2222, 1996.
128. Männikkö, M., Lenkkeri, U., Kashtan, C.E., Kestilä, M., Holmberg, C., and Tryggvason, K.: Haplotype analysis of congenital nephrotic syndrome of the Finnish type in non-Finnish families. *J. Am. Soc. Nephrol.* 7(12) 2700-2703, 1996.
129. Zhang, X., Vuolteenaho, R., and Tryggvason, K.: Structure of the human laminin  $\alpha 2$ -chain gene (*LAMA2*) which is affected in congenital muscular dystrophy. *J. Biol. Chem.*, 271, 27664-27669, 1996.
130. Soini, Y., Määttä, M., Salo, S., Tryggvason, K., and Autio-Harmainen: Expression of the laminin  $\gamma 2$  chain in pancreatic adenocarcinomas. *H. J. Pathol.*, 180, 290-294, 1996.
131. Groffen, A.J.A., Buskens, C.A.F., Tryggvason, K., Veerkamp, J.H., Monnens, L.H. and van den Heuvel, L.P.W.: Expression and functional characterization of human perlecan domains I and II synthesized by baculovirus-infected insect cell. *Eur. J. Biochem.* 241, 827-834, 1996.
132. Allamand, V., Sunada, Y., Salih, M.A.M., Straub, V., Ozo, C.O., Al-Turaike, M.H.S., Akbar, M., Kolo, T., Xhang, X., Yurchenco, P.D., Tryggvason, K., and Campbell, K.P. Mild congenital muscular dystrophy in two patients with an internally deleted laminin alpha-2 chain. *Hum. Mol. Genet.* 6:747-752, 1997.
133. Guicheney, P., Vignier, N., Helbling-Leclerc, A., Nissinen, M., Zhang, X., Cruaud, C., Lambert, J.C., Richelme, C., Topaloglu, H., Merlini, L., Barois, A., Schwartz, K., Tomé, F.M.S., Tryggvason, K., and Fardeau, M. Genetics of laminin  $\alpha 2$  chain (or merosin) deficient congenital muscular dystrophy: from identification of mutations to prenatal diagnosis. *Neuromusc. Disord.* 7: 180-186, 1997.
134. Männikkö, M., Kestilä, M., Lenkkeri, U., Alakurtti, H., Holmberg, C., Leisti, J., Salonen, R., Aula, P., Mustonen, A. Peltonen, L., and Tryggvason, K.: Improved prenatal diagnosis of the congenital nephrotic syndrome of the Finnish type based on DNA analysis. *Kidney Int.* 51: 868-872, 1997.
135. Pulkkinen, L., McGrath, J., Airenne, T., Haakana, H., Tryggvason, K., Kivirikko, S., Meneguzzi, G., Christiano, A., and Uitto, J.: Detection of *LAMC2* mutations Herlitz junctional epidermolysis bullosa. *Mol. Med.*, 3, 124-135, 1997.
136. Groffen, A.J., Hop, F.W., Tryggvason, K., Dijkman, H., Assman, K.J., Veerkamp, J.H., Monnens, L.A., and Van den Heuvel, L.P. Evidence for the existence of multiple heparan sulfate proteoglycans in the human glomerular basement membrane and mesangial matrix. *Eur. J. Biochem*, 247, 175-182, 1997.
137. Iivanainen, A., Korttesmaa, J., Sahlberg, C., Morita, T., Bergmann, U., Thesleff, I., and Tryggvason, K. Primary structure, developmental expression and immunolocalization of the murine laminin  $\alpha 4$  chain. *J. Biol. Chem.*, 272: 27862-27868, 1997.

138. Kainulainen T., Autio-Harmainen, H., Oikarinen, A., Salo, S., Tryggvason, K. and Salo, T.: Altered distribution and synthesis of laminin-5 (kalinin) in oral lichen planus, epithelial dysplasias and squamous cell carcinomas. *Br. J. Dermatol.*, 136, 331-336, 1997.
139. van der Laan, L.J., Kangas, M., Dopp, E.A., Broug-Holub, E., Elomaa, O., Tryggvason, K., and Kraal, G.: Macrophage scavenger receptor MARCO: in vitro and in vivo regulation and involvement in the anti-bacterial host defense. *Immunol. Lett.* 57, 203-208, 1997.
140. Dehan, P., Walteregny, D., Beschin, A., Noel, A., Castronovo, V., Tryggvason, K., Deleval, J., and Foidart, J.-M.: Loss of type IV collagen alpha-5 and alpha-6 chains in human invasive prostate carcinomas. *Am. J. Pathol.* 151, 1097-1104, 1997.
141. Lenkkeri, U., Kestilä, M., Lamerdin, J., McCready, P., Adamson, A., Olsen, A. and Tryggvason, K.: Structure of the human amyloid precursor-like protein gene APLP1 at 19q13.1. *Hum. Genet.* 102, 192-196, 1998.
142. Guicheney, P., Vignier, N., Zhang, X., He, Y., Cruaud, C., Frey, V., Helblinc-Leclerc, A., Richard, P., Estournet, B., Merlini, L., Topaloglu, H., Mora, M., Harpey, J.-P., Haenggeli, C.-A., Barois, A., Hainque, B., Schwartz, K., Tomé, F., Fardeau, M. and Tryggvason, K.: PCR-based mutation screening of the laminin  $\alpha 2$  chain gene (LAMA2): application to prenatal diagnosis and search of founder effects in congenital muscular dystrophy. *J. Med. Genet.* 35, 211-217, 1998.
143. Elomaa, O., Sankala, M., Pikkarainen, T., Bergmann, U., Tuuttila, A. Raatikainen, -Ahokas, A., Sariola, H., and Tryggvason, K.: Structure of the human macrophage receptor and characterization of its bacacteria-binding domain. *J. Biol. Chem.* 273, 4530-4538, 1998.
144. Kestilä, M., Lenkkeri, U., Männikkö, M., Lamerdin, J., McCready, P., Putaala, H., Ruotsalainen, V., Morita, T., Nissinen, M., Herva, R., Kashtan, C.E., Peltonen, L., Holmberg, C., Olsen, A., and Tryggvason, K.: Positionally cloned gene for a novel glomerular protein - nephrin - is mutated in congenital nephrotic syndrome. *Mol. Cell*, 1, 575-582, 1998.
145. Martin, P., Heiskari, N., Zhou, J., Leinonen, A., Tumelius, T., Hertz, J. M., Barker, D., Gregory, M., Atkin, C., Styrkársdóttir, U., Neumann, H., Springate, J., Shows, T., Pettersson, E., and Tryggvason, K.: High mutation detection rate in the COL4A5 collagen gene in suspected Alport syndrome using PCR and direct DNA sequencing. *J. Am. Soc. Nephrol.* 9, 2291-2301, 1998.
146. Tuuttila, A., Morgunova, E., Bergmann, U., Lindqvist, Y., Maskos, K., Fernandez-Catalan, C., Bode, W., Tryggvason, K. and Schneider, G.: Three-dimensional structure of free human inhibitor of metalloproteinases-2 at 2.1 Å resolution. *J. Mol. Biol.* 284, 1133-1140, 1998.
147. Van der Laan, L.J.W., Dopp, E.A., Haworth, R., Pikkarainen, T. Kangas, M. Elomaa, O. Dijkstra, C.D. Gordon, S. Tryggvason, K. Kraal, G.: Regulation and functional involvement of macrophage scavenger receptor MARCO in clearance of bacteria in vivo. *J. Immunol.* 162, 939-947, 1999.
148. Munaut, C., Salonurmi, T., Kontusaari, S., Reponen, P., Morita, T., Foidart, J.-M., and Tryggvason, K.: Murine matrix metalloproteinase 9 gene: 5' upstream region contains cis-acting elements for expression in osteoclasts and migrating keratinocytes in transgenic mice. *J. Biol Chem.* 274, 5588-5596, 1999.
149. Lenkkeri, U., Männikkö, M., McCready, P., Lamerdin, J., Gribouval, O., Niaudet, P., Antignac, C., Kashtan, C. E., Holmberg, C., Olsen, A., Kestilä, M., and Tryggvason, K.: Structure of the gene for congenital nephrotic syndrome of the Finnish type (*NPHS1*) and characterization of mutations. *Am. J. Hum. Genet.* 64, 51-61, 1999.
150. Iivanainen, A., Morita, T., and Tryggvason, K.: Molecular cloning and tissue specific expression of a novel murine laminin  $\gamma 3$  chain. *J. Biol. Chem.*, 274, 14107-14111, 1999.

151. Kangas, M., Brännström, A., Elomaa, O., Matsuda, Y., Eddy, R., Shows, T. B., and Tryggvason, K.: Structure and chromosomal localization of the human and murine genes for the macrophage MARCO receptor. *Genomics* 58, 82-89, 1999.
152. Pikkarainen, T., Brännström, A., and Tryggvason, K.: Expression of macrophage MARCO receptor induces formation of dendritic plasma membrane processes. *J. Biol. Chem.* 274, 10975-10982, 1999.
153. Salo, S., Haakana, H., Kontusaari, S., Hujanen, E., Kallunki, T., and Tryggvason, K.: Laminin-5 promotes adhesion and migration of epithelial cells: identification of a migration-related element in the  $\gamma 2$  chain gene (LAMC2) with activity in transgenic mice. *Matrix Biol.* 18, 197-210, 1999.
154. Morgunova, E., Tuuttila, A., Bergmann, U., U., Isupov, M., Lindqvist, Y., Schneider, G., and Tryggvason, K.: Structure of human pro-matrix metalloproteinase-2: activation mechanism revealed, *Science* 284, 1667-1670, 1999.
155. Palecanda, A., Paulauskis, J., Al-Mutari, E., Imrich, A., Qin, G., Suzuki, H., Kodama, T., Tryggvason, K., Koziel, H., and Kobzik, L.: Role of scavenger receptor MARCO in alveolar binding of unopsonized environmental particles. *J. Exp. Med.* 189, 1497-1506, 1999.
156. Ruotsalainen, V., Ljungberg, P., Wartiovaara, J., Lenkkeri, U., Kestilä, M., Jalanko, H., Holmberg, C., and Tryggvason, K.: Nephlin is located in the slit membrane of glomerular podocytes. *Proc. Natl. Acad. Sci., USA* 96, 7962-7967, 1999.
157. Määttä, M., Soini, Y., Pääkkö, P., Salo, S., Tryggvason, K., and Autio-Harmainen, H.: Expression of the laminin  $\gamma 2$  chain in different histological types of lung carcinoma. A study by immunohistochemistry and in situ hybridization. *J. Pathol.* 188, 361-368, 1999.
158. Skyldberg, B., Salo, S., Eriksson, E., Aspenblad, U., Moberger, B., Tryggvason, K. and Auer, G.: Laminin-5 as a marker of invasiveness in cervical lesions. *J. Natl. Cancer Inst.* 91, 1882-1887, 1999.
159. Ito, S., Naito, M., Kobayashi, Y., Takatsuka, H., Jiang, S., Usuda, H., Umezu, H., Hasegawa, G., Arakawa, M., Shultz, L.D., Elomaa, O. and Tryggvason, K.: Roles of a macrophage receptor with collanenous structure (MARCO) in host defense and heterogeneity of splenic marginal zone macrophages. *Arch. Histol. Cytol.* 62, 83-95, 1999.
160. Sahlberg, C., Reponen, P., Tryggvason, K. and Thesleff, I.: Timp-1, -2 and -3 show coexpression with gelatinases A and B during mouse tooth morphogenesis. *Eur. J. Oral Sci.* 107, 121-130, 1999.
161. Geberhiwot, T., Ingerpuu, S., Pedraza, C., Neira, M., Lehto, U., Virtanen, I., Kortessmaa, J., Tryggvason, K., Engvall, E. and Patarroyo, M.: Blood platelets contain and secrete laminin-8 ( $\alpha 4:\beta 1:\gamma 1$ ) and adhere to laminin-8 via  $\alpha 6:\beta 1$  integrin. *Exp. Cell Res.* 253, 723-32, 1999.
162. Parpala-Spärman, T., Lukkarinen, O., Heikkilä, P. and Tryggvason, K.: A novel surgical organ perfusion method for effective ex and in vivo gene transfer into renal glomerular cells. *Urol. Res.* 27, 97-102, 1999.
163. Putaala, H., Sainio, K., Sariola, H. and Tryggvason, K.: Primary structure of mouse and rat nephlin cDNA and structure and developmental expression of the mouse gene. *J. Am. Soc. Nephrol.* 11, 991-1001, 2000
164. Kortessmaa, J., Yurchenco, P., and Tryggvason, K.: Recombinant laminin-8 ( $\alpha 4:\beta 1:\gamma 1$ ): production, purification and interactions with integrins. *J. Biol. Chem.*, 275, 14853-14859, 2000.
165. Airene, T., Lin, Y., Olsson, M., Ekblom, P., Vainio, S. and Tryggvason, K.: Differential expression of mouse laminin  $\gamma 2$  and  $\gamma 2^*$  chain transcripts. *Cell Tissue Res.*, 300, 129-138, 2000.
166. Zhou, Z., Apte, S.S., Soininen, R., Cao, R., Baaklini, G.Y., Rauser, R.W., Wang, J., Cao, Y. and Tryggvason, K.: Impaired endochondral ossification and

- angiogenesis in mice deficient in membrane type matrix metalloproteinase I (MT1-MMP), *Proc. Natl. Acad. Sci., USA* 97, 4052-4057, 2000.
167. Jais, J.P., Knebelmann, B., Giatras, I., de Marchi, M., Rizzoni, G., Renieri, A., Weber, M., Gross, O., Netzer, K-O., Flinter, F., Pirson, Y., Verellen, C., Wieslander, J., Persson, U., Tryggvason, K., Martin, P., Hertz, J.M., Schröder, C., Sanak, M., Krejcova, S., Carvalho, M.F., Saus, J., Antignac, C., Smeets, H. and Gubler, M. C.: X-linked Alport syndrome: natural history in 195 families and genotype- phenotype correlations in males. *J. Am. Soc. Nephrol.* 11, 649-657, 2000.
  168. Martin, P., Heiskari, N., Pajari, H., Grönhagen-Riska, C., Kääriäinen, H., Koskimies, O. and Tryggvason, K.: Spectrum of COL4A5 mutations in Finnish Alport syndrome patients. *Hum. Mut.* 15, 579, 2000.
  169. Geberhiwot, T., Ingerpuu, P., Pedraza, C., Neira, M., Virtanen, I., Tryggvason, K. and Patarroyo, M.: Erythromegakaryocytic cells synthesize laminin-8 ( $\alpha 4\beta 1\gamma 1$ ). *Exp. Cell Res.* 254, 189-195, 2000.
  170. Li, C., Ruotsalainen, V., Tryggvason, K., Shaw, A.S. and Miner, J.H.: CD2AP is expressed with nephrin in developing podocytes and is found widely in mature kidney and elsewhere. *Am. J. Physiol. Renal Physiol.* 279, 785-792, 2000.
  171. Geberhiwot, T., Wondimu, Z., Salo, Sirpa., Pikkarainen, T., Korttesmaa, J., Tryggvason, K., Virtanen, I. and Patarroyo, M.: Chain specificity assignment of monoclonal antibodies to human laminins by using recombinant laminin  $\beta 1$  and  $\gamma 1$  chains. *Matrix Biol.*, 19, 163-167, 2000.
  172. Patrakka, J., Kestilä, M., Wartiovaara, J., Ruotsalainen, V., Tissari, P., Lenkkeri, U., Männikkö, M., Visapää, I., Holmberg, C., Rapola, J., Tryggvason, K. and Jalanko, H.: Congenital nephrotic syndrome (NPHS1): Features resulting from different mutations in Finnish patients. *Kidney Int.* 58, 972-980, 2000.
  173. Ruotsalainen, V., Patrakka, J., Tissari, P., Hess, M., Kestilä, M., Holmberg, C., Salonen, R., Heikinheimo, M., Wartiovaara, J., Tryggvason, K. and Jalanko, H.: Role of nephrin in cell junction formation in human nephrogenesis. *Am. J. Pathol.* 157, 1905-1916, 2000.
  174. Pedraza, C., Geberhiwot, T., Ingerpuu, S., Assefa, D., Wondimu, Z., Korttesmaa, K., Tryggvason, K., Virtanen, I. and Patarroyo, M.: Monocytic cells synthesize, adhere to, and migrate on laminin-8 ( $\alpha 4\beta 1\gamma 1$ ). *J. Immunol.* 165, 5831-5838, 2000.
  175. Putaala, H., Soininen, R., Kilpeläinen, P., Wartiovaara, J. and Tryggvason, K.: The murine nephrin gene is specifically expressed in kidney, brain and pancreas. Inactivation of the gene leads to massive proteinuria and neonatal death. *Hum. Mol. Genet.* 10, 1-8, 2001.
  176. Patrakka, J., Ruotsalainen, V., Qvist, E., Laine, J., Holmberg, C., Tryggvason, K. and Jalanko, H.: Nephrin and recurrence of nephrotic syndrome in patients with congenital nephrotic syndrome. *J. Am. Soc. Nephrol.* 12, 289-296, 2001.
  177. Martin, P. and Tryggvason, K.: Two novel alternatively spliced 9-bp exons in the COL4A5 gene. *Ped. Nephrol.* 16, 41-44, 2001.
  178. Geberhiwot, T., Assefa, D., Korttesmaa, J., Ingerpuu, S., Pedraza, C., Wondimu, Z., Charo, J., Kiessling, R., Virtanen, I., Tryggvason, K. and Patarroyo, M.: Laminin-8 ( $\alpha 4\beta 1\gamma 1$ ) is synthesized by lymphoid cells, promotes lymphocyte migration and costimulates T cell proliferation. *J. Cell Sci.* 114, 423-433, 2001.
  179. Patrakka, J., Ruotsalainen, V., Ketola, I., Holberg, C., Tryggvason, K. and Jalanko, H.: Expression of nephrin in pediatric kidney diseases. *J. Am. Soc. Nephrol.*, 12, 289-296, 2001.
  180. Beltcheva, O., Martin, P., Lenkkeri, U. and Tryggvason, K.: Mutation spectrum in the nephrin gene (NPHS1) in congenital nephrotic syndrome. *Hum. Mut.*, 17, 368-373, 2001.
  181. Doublie, S., Ruotsalainen, V., Salvidio, G., Lupia, E., Biancone, L., Conaldi, P.G., Reponen, P., Tryggvason, K. and Camussi, G.: Nephrin redistribution on

- podocytes is a potential pathomechanism for proteinuria in patients with primary acquired nephrotic syndrome. *Am. J. Pathol.* 158, 1723-1731, 2001.
182. Heikkilä, P., Tibell, A., Morita, T., Chen, Y., Wu, G., Sado, Y., Ninomiya, Y., Pettersson, E. and Tryggvason, K.: Adenovirus-mediated transfer of type IV collagen  $\alpha 5$  chain cDNA into swine kidney in vivo: deposition of the protein into the glomerular basement membrane. *Gene Therapy*, 8, 882-890, 2001.
  183. Patton, B.L., Cunningham, J.M., Thyboll, J., Korttesmaa, J., Westerblad, H., Edström, L., Tryggvason, K., Sanes, J.R.: Properly formed but improperly localized synaptic specializations in the absence of laminin  $\alpha 4$ . *Nature Neurosci.* 4, 597-604, 2001.
  184. Parpala-Spärman, T., Pääkkö, P., Kortteinen, P., Salonurmi, T., Lukkarinen, O. and Tryggvason, K.: Closed-circuit organ perfusion technique for gene transfer into the lungs. An experimental trial on farm pigs. *Eur. J. Clin. Invest.* 31, 264-271, 2001.
  185. Lenander, C., Habermann, J.K., Öst, Å., Nilsson, B., Schimmelpenning, H., Tryggvason, K. and Auer, G.: Laminin-5  $\gamma 2$  chain expression correlates with unfavorable prognosis in colon carcinomas. *Anal. Cell. Pathol.* 22, 201-209, 2001.
  186. Habermann, J., Lenander, C., Roblick, U.J., Krüger, S., Ludwig, D., Alaiya, A., Freitag, S., Drümbgen, L., Bruch, H-P., Stange, E., Salo, S., Tryggvason, K., Auer, G. and Schimmelpenning, H.: Ulcerative colitis and colorectal carcinoma. DNA-profile, laminin-5  $\gamma 2$  chain and cyclin a expression as early markers for risk assessment. *Scand. J. Gastroenterol.* 36, 751-758, 2001.
  187. Liu, L., Cotta Doné, S., Khoshnoodi, J., Bertorello, A., Wartiovaara, J., Berggren, P-O. and Tryggvason, K.: Defective nephrin trafficking caused by missense mutations in the *NPHS1* gene: insight into the mechanisms of congenital nephrotic syndrome. *Hum. Mol. Genet.* 10, 2637-2644, 2001.
  188. Srivastava, T., Whiting, J.M., Garola, R.E., Dasouki, M.J., Ruotsalainen, V., Tryggvason, K., Hamed, R., and Alon, U.S.: Podocyte proteins in Galloway-Mowat syndrome. *Pediatr. Nephrol.* 16, 1022-1029, 2001.
  189. Doi, M., Thyboll, J., Korttesmaa, J., Jansson, K., Iivanainen, A., Parvardeh, M., Timpl, R., Hedin, U., Swedenborg, J. and Tryggvason K.: Recombinant human laminin-10 ( $\alpha 5\beta 1\gamma 1$ ): Production, purification and migration promoting activity on vascular endothelial cells. *J. Biol. Chem.*, 277, 12741-12748, 2002.
  190. Thyboll, J., Korttesmaa, J., Cao, R., Soininen, R., Wang, L., Iivanainen, A., Sorokin, L., Risling, M., Cao, Y. and Tryggvason, K.: Deletion of the laminin  $\alpha 4$  chain leads to impaired microvessel maturation. *Mol. Cell. Biol.*, 22, 1194-1202, 2002.
  191. Koziell, A., Grech, V., Hussain, S., Lee, G., Lenkkeri, U., Tryggvason, K. and Scambler, P.: Genotype/phenotype correlations of *NPHS1* and *NPHS2* mutations in nephrotic syndrome advocate a functional interrelationship in glomerular filtration. *Hum Mol Genet*, 15, 379-388, 2002.
  192. Nordström, B., Einhorn, N., Silfversward, C., Sjövall, K., Tryggvason, K., and Auer, K.: Laminin-5 gamma-2 chain as an invasivity marker for uni- and multifocal lesions in the lower anogenital tract. *Int. J. Cancer*, 13, 105-109, 2002.
  193. Patrakka, J., Ruotsalainen, V., Reponen, P., Qvist, E., Laine, J., Holmberg, C., Tryggvason, K. and Jalanko, H.: Recurrence of nephrotic syndrome in kidney grafts of *NPHS1* patients: Role of nephrin. *Transplantation*, 15, 394-403, 2002.
  194. Huh, W., Kim, M.K., Kim, Y.G., Oh, H.Y., Ruotsalainen, V., and Tryggvason, K.: Expression of nephrin in acquired human glomerular disease. *Nephrol. Dial. Transplant.*, 17, 478-484, 2002.
  195. Brännström, A., Sankala, M., Tryggvason, K., and Pikkarainen, T.: Arginine residues in domain V have a central role for bacterial binding activity of macrophage scavenger receptor MARCO. *Biochem. Biophys. Res. Commun.*, 290, 1462-1469, 2002.

196. Patrakka, J., Martin, P., Salonen, R., Ruotsalainen, V., Kestilä, M., Männikkö, M., Ryyänen, M., Rapola, J., Holmberg, C., Tryggvason, K. and Jalanko, H.: Proteinuria and prenatal diagnosis of congenital nephrosis in fetal carriers of nephrin gene mutations. *Lancet*, 359, 1575-1577, 2002.
197. Pendas, A., Zhou, Z., Cadinanos, J., Freije, J. M., Wang, J., Hultenby, K., Astudillo, A., Wernerson, A., Rodriguez, F., Tryggvason, K., and Lopez-Otin, C.: Defective prelamin A processing and muscular and adipocyte alterations in Zmpste24 metalloproteinase deficient mice. *Nature Genet.*, 31, 94-99, 2002.
198. Smirnov, S.P., McDearmon, E.L., Li, S., Ervasti, J.M., Tryggvason, K., and Yurchenco, P.D: Contribution of the LG modules and furin-processing to laminin-2 functions. *J. Biol. Chem.*, 277, 18928-18937, 2002.
199. Morgunova, E., Tuuttila, A., Bergmann, U., and Tryggvason, K.: Structural insight into the complex formation of latent matrix metalloproteinase 2 with tissue inhibitor of metalloproteinase 2. *Proc. Natl. Acad. Sci. USA*, 99, 7414-7419, 2002.
200. Yan, K., Khoshnoodi, J., Ruotsalainen, V., and Tryggvason, K.: N-linked glycosylation is critical for the plasma membrane localization of nephrin. *J. Am. Soc. Nephrol.* 13, 1385-1389, 2002.
201. Sankala, M., Brännström, A., Schultess, T., Bergmann, U., Morgunova, E., Engel, J., Tryggvason, K. and Pikkarainen, T.: Characterization of recombinant soluble macrophage scavenger receptor MARCO. *J. Biol. Chem.* 277, 33378-33385, 2002..
202. Petäjänieniemi, N., Korhonen, M., Korttesmaa, J., Tryggvason, K., Sekiguchi, K., Fujiwara, H., Sorokin, L., Thornell, L.E., Assefa, D., Patarroyo, M., Virtanen, I.: Localization of laminin alpha4-chain in developing and adult human tissues. *J Histochem. Cytochem.* 50, 1113-1130, 2002.
203. Franzke, C.W., Tasanen, K., Schacke, H., Zhou, Z., Tryggvason, K., Mauch, C., Zigrino, P., Sunnarborg, S., Lee, D.C., Fahrenholz, F., Bruckner-Tuderman, L.: Transmembrane collagen XVII, an epithelial adhesion protein, is shed from the cell surface by ADAMs. *EMBO J.* 21, 5026-5035, 2002.
204. Saleem, M.A., Ni, L., Witherden, I., Tryggvason, K., Ruotsalainen, V., Mundel, P., Mathieson, P.W.: Co-localization of nephrin, podocin, and the actin cytoskeleton: evidence for a role in podocyte foot process formation. *Am. J. Pathol.* 161, 1459-1466, 2002.
205. Korttesmaa, J., Doi, M., Patarroyo, M., Tryggvason, K.: Chondroitin sulphate modification in the alpha4 chain of human recombinant laminin-8 (alpha4beta1gamma1). *Matrix Biol.* 21, 483-486, 2002.
206. Gu, Y.C., Korttesmaa, J., Tryggvason, K., Persson, J. Ekblom, P., Jacobsen, S.E., Ekblom, M.: Laminin isoform-specific promotion of adhesion and migration of human bone marrow progenitor cells. *Blood*, 101, 877-885, 2003.
207. Wernerson, A., Dunér, F., Pettersson, E., Widholm, S., Berg, U., Ruotsalainen, V., Tryggvason, K., Hultenby, K. and Söderberg, M.: Reduced nephrin expression in glomeruli of patients with minimal change nephrotic syndrome. *Nephrol. Dial Transplant.* 18, 70-76, 2003.
208. Rossi, M. Morita, H., Sormunen, R., Airene, S., Kreivi, M., Wang, L., Fukai, N., Olsen, B.R., Tryggvason, K., Soininen, R.: Heparan sulfate chains of perlecan are indispensable in the lens capsule but not in the kidney. *EMBO J.* 22, 236-245, 2003.
209. Beltcheva, O., Kontusaari, S., Fetissov, S., Putaala, H., Kilpeläinen, P., Hökfelt, T., and Tryggvason, K.: Alternatively used promoters and distinct elements direct tissue-specific expression of nephrin. *J. Am. Soc. Nephrol.* 4, 352-358, 2003.
210. Adair-Kirk, T.L., Atkinson, J.J., Broekelmann, T.J., Doi, M., Tryggvason, K., Miner, J.H., Mecham, R.P., and Senior, R.M.: A site on laminin alpha5, AQARSAASKVKVSMKF, induces inflammatory cell production of matrix metalloproteinase-9 and chemotaxis. *J Immunol.* 171, 398-406, 2003

211. Lundgren, C., Frankendal, B., Silfversward, C., Nilsson, B., Tryggvason, K., Auer, G., and Nordstrom, B.: Laminin-5 gamma2-chain expression and DNA ploidy as predictors of prognosis in endometrial carcinoma. *Med Oncol.* 20, 147-156, 2003.
212. Lahdenpera, J., Kilpelainen, P., Liu, X.L., Pikkarainen, T., Reponen, P., Ruotsalainen, V., and Tryggvason, K.: Clustering-induced tyrosine phosphorylation of nephrin by Src family kinases. *Kidney Int.* 64, 404-413, 2003.
213. Khoshnoodi, J., Sigmundsson, K., Ofverstedt, LG, Skoglund, U, Obrink, B, Wartiovaara, J. and Tryggvason, K.: Nephrin Promotes Cell-Cell Adhesion through Homophilic Interactions. *Am J Pathol*, 163, 2337-2346, 2003.
214. Jais, JP., Knebelmann, B., Giatras, I., De Marchi, M., Rizzoni, G., Renieri, A., Weber, M., Gross, O., Netzer, KO., Flinter, F., Pirson, Y., Dahan, K., Wieslander, J., Persson, U., Tryggvason, K., Martin, P., Hertz, JM., Schroder, C., Sanak, M., Carvalho, MF., Saus, J., Antignac, C., Smeets, H., Gubler, MC.: X-linked alport syndrome: natural history and genotype-phenotype correlations in girls and women belonging to 195 families: a "European Community Alport Syndrome Concerted Action" study. *J Am Soc Nephrol.* 14, 2603-10. 2003.
215. Bartlett, JD., Zhou, Z., Skobe, Z., Dobeck, JM. and Tryggvason, K.: Delayed tooth eruption in membrane type-1 matrix metalloproteinase deficient mice. *Connect Tissue Res.* 44, 300-304, 2003.
216. Lenander, C., Roblick, U.J., Habermann, JK.K., Ost, A., Tryggvason, K., and Auer, G.: Laminin-5 gamma-2 chain expression: a marker of early invasiveness in colorectal adenomas. *Mol. Pathol.* 56, 342-346, 2003.
217. Salonurmi, T., Parikka, M., Kontusaari, S., Pirila, E., Munaut, C., Salo, T. and Tryggvason, K.: Overexpression of TIMP-1 under the MMP-9 promoter interferes with wound healing in transgenic mice. *Cell Tissue Res.* 315, 27-37, 2004.
218. Tran, P.K., Tran-Lundmark, K., Soininen, R., Tryggvason, K., Thyberg, J., and Hedin, U.: Increased intimal hyperplasia and smooth muscle cell proliferation in transgenic mice with heparan sulfate deficient perlecan. *Circ. Res.* 94, 550-558, 2004.
219. Kuusiniemi, A.M., Kestilä, M., Patrakka, J., Lahdenkari, A.T., Ruotsalainen, V., Holmberg, C., Karikoski, I., Salonen, R., Tryggvason, K., and Jalanko, H.: Tissue expression of nephrin in human and pig. *Pediatr. Res.* 55, 774-781, 2004.
220. Ruotsalainen, V., Reponen, P., Khoshnoodi, J., Kilpeläinen, P., and Tryggvason, K.: Monoclonal antibodies to human nephrin. *Hybrid Hybridomics*, 23, 55-63, 2004.
221. Liu, X.L., Done, S.C., Yan, K., Kilpelainen, P., Pikkarainen, T., Tryggvason, K.: Defective trafficking of nephrin missense mutants rescued by a chemical chaperone. *J Am Soc Nephrol.* 15, 1731-1738, 2004.
222. Zhou, Z., Doi, M., Wang, J., Cao, R., Liu, B., Chan, K.M., Kortessmaa, J., Sorokin, L., Cao, Y., Tryggvason, K.: Deletion of laminin-8 results in increased tumor neovascularization and metastasis in mice. *Cancer Res.* 64, 4059-4063, 2004.
223. Zhou, Z., Wang, J., Cao, R., Morita, H., Soininen, R., Chan, K.M., Liu, B., Cao, Y., Tryggvason, K.: Impaired angiogenesis, delayed wound healing and retarded tumor growth in perlecan heparan sulfate-deficient mice. *Cancer Res.* 64, 4699-4702, 2004.
224. Arredouani, M., Yang, Z., Ning, Y., Qin, G., Soininen, R., Tryggvason, K., Kobzik, L.: The scavenger receptor MARCO is required for lung defense against pneumococcal pneumonia and inhaled particles. *J Exp Med.* 200, 267-272, 2004.
225. Mirastschijski, U., Zhou, Z., Rollman, O., Tryggvason, K., Agren, M.S.: Wound healing in membrane-type-1 matrix metalloproteinase-deficient mice. *J Invest Dermatol.* 123, 600-602, 2004

226. Stoltzfus, P., Salo, S., Eriksson, E., Aspenblad, U., Tryggvason, K., Auer, G., Avall-Lundqvist E.: Laminin-5 gamma2 chain expression facilitates detection of invasive squamous cell carcinoma of the uterine cervix. *Int J Gynecol Pathol.* 23, 215-222, 2004.
227. Wondimu, Z., Geberhiwot, T., Ingerpuu, S., Juronen, E., Xie, X., Lindbom, L., Doi, M., Korttesmaa, J., Thyboll, J., Tryggvason, K., Fadeel, B., Patarroyo, M.: An endothelial laminin isoform, laminin 8 (alpha-4:beta-1:gamma-1), is secreted by blood neutrophils, promotes neutrophil migration and extravasation, and protects neutrophils from apoptosis. *Blood* 15, 1859-1866, 2004.
228. Byrne, L.C., Zhou, Z., Tryggvason, K., Hokfelt, T., and Fetissov, S.O.: Altered NPY and AgRP in membrane type-1 matrix metalloproteinase-deficient mice. *Neuroreport.* 15, 569-74, 2004.
229. Nishibori, Y., Liu, L., Hosoyamada, M., Endou, H., Kudo, A., Takenaka, H., Higashihara, E., Bessho, F., Takahashi, S., Kershaw, D., Ruotsalainen, V., Tryggvason, K., Khoshnoodi, J., and Yan, K.: Disease-causing missense mutations in NPHS2 gene alter normal nephrin trafficking to the plasma membrane. *Kidney Int.* 66, 1755-1765, 2004.
230. Wartiovaara, J., Öfverstedt, L-G., Khoshnoodi, J., Zhang, J., Mäkelä, E., Sandin, S., Ruotsalainen, V., Cheng, R.H., Jalanko, J., Skoglund, U., and Tryggvason, K.: Nephrin strands contribute to a porous slit diaphragm scaffold as revealed by electron tomography. *J. Clin. Invest.* 114, 1475-1483, 2004.
231. Liu, X.L., Kilpelainen, P., Hellman, U., Sun, Y., Wartiovaara, J., Morgunova, E., Pikkarainen, T., Yan, K., Jonsson, A.P., and Tryggvason, K.: Characterization of the interactions of the nephrin intracellular domain. *FEBS J.* 272, 228-243, 2005.
232. Stoltzfus, P., Heselmeyer-Haddad, K., Castro, J., White, N., Silfversward, C., Sjøvall, K., Einhorn, N., Tryggvason, K., Auer, G., Ried, T., and Nordstrom, B.: Gain of chromosome 3q is an early and consistent genetic aberration in carcinomas of the vulva. *Int J Gynecol Cancer* 15, 120-126, 2005.
233. Melles, E., Jornvall, H., Tryggvason, S., Danielsson, K.G., Ekberg, K., Tryggvason, K., Wahren, J., and Bergman, T.: Degradation of proinsulin C-peptide in kidney and placenta extracts by a specific endoprotease activity. *Cell Mol Life Sci.* 61, 2979-2982, 2005.
234. Oblander, S.A., Zhou, Z., Galvez, B.G., Starcher, B., Shannon, J.M., Durbeej, M., Arroyo, A.G., Tryggvason, K., and Apte, S.S.: Distinctive functions of membrane type 1 matrix-metalloproteinase (MT1-MMP or MMP-14) in lung and submandibular gland development are independent of its role in pro-MMP-2 activation. *Dev Biol.* 277, 255-269, 2005.
235. Galvez, B.G., Genis, L., Matias-Roman, S., Oblander, S.A., Tryggvason, K., Apte, S.S., and Arroyo, A.G.: Membrane type 1-matrix metalloproteinase is regulated by chemokines monocyte-chemoattractant protein-1/ccl2 and interleukin-8/CXCL8 in endothelial cells during angiogenesis. *J Biol Chem.* 280, 1292-1298, 2005.
236. Wallquist, W., Plantman, S., Thams, S., Thyboll, J., Korttesmaa, J., Lannergren, J., Domogatskaya, A., Ogren, S.O., Risling, M., Hammarberg, H., Tryggvason, K., Cullheim, S.: Impeded interaction between Schwann cells and axons in the absence of laminin alpha4. *J Neurosci.* 25, 3692-3700, 2005.
237. Morita, H., Yoshimura, A., Inui, K., Ideura, T., Watanabe, H., Wang, L., Soininen, R., Tryggvason, K.: Heparan sulfate of perlecan is involved in glomerular filtration. *J Am Soc Nephrol.* 16, 1703-1710, 2005.
238. Liu, B., Wang, J., Chan, K.M., Tjia, W.M., Deng, W., Guan, X., Huang, J.D., Li, K.M., Chau, P.Y., Chen, D.J., Pei, D., Pendas, A.M., Cadinanos, J., Lopez-Otin, C., Tse, H.F., Hutchison, C., Chen, J., Cao, Y., Cheah, K.S., Tryggvason, K., Zhou, Z. Genomic instability in laminopathy-based premature aging. *Nature Med.* 11, 780-785, 2005.
239. Varela, I., Cadinanos, J., Pendas, A.M., Gutierrez-Fernandez, A., Folgueras, A.R., Sanchez, L.M., Zhou, Z., Rodriguez, F.J., Stewart, C.L., Vega, J.A.,



- Tryggvason, K., Freije, J.M., Lopez-Otin, C. Accelerated ageing in mice deficient in Zmpste24 protease is linked to p53 signalling activation. *Nature* 437:564-568, 2005
240. Wang J, Hoshijima M, Lam J, Zhou Z, Jokiel A, Dalton ND, Hultenby K, Ruiz-Lozano P, Ross J Jr, Tryggvason K, Chien KR.: Cardiomyopathy associated with microcirculation dysfunction in laminin alpha4 chain-deficient mice. *J Biol Chem.* 281:213-220, 2006
241. Arredouani MS, Palecanda A, Koziel H, Huang YC, Imrich A, Sulahian TH, Ning YY, Yang Z, Pikkarainen T, Sankala M, Vargas SO, Takeya M, Tryggvason K, Kobzik L.: MARCO is the major binding receptor for unopsonized particles and bacteria on human alveolar macrophages. *J Immunol.* 175:6058-6064, 2005
242. Chen Y, Pikkarainen T, Elomaa O, Soininen R, Kodama T, Kraal G, Tryggvason K.: Defective microarchitecture of the spleen marginal zone and impaired response to a thymus-independent type 2 antigen in mice lacking scavenger receptors MARCO and SR-A. *J Immunol.* 175:8173-8180, 2005.
243. Qian H, Tryggvason K, Jacobsen SE, Ekblom M.: Contribution of  $\alpha$ 6-integrins to hematopoietic stem and progenitor cell homing to bone marrow and collaboration with  $\alpha$ 4-integrins. *Blood* 107, 3503-3510, 2006
244. Martens JH, Kzhyshkowska J, Falkowski-Hansen M, Schledzewski K, Gratchev A, Mansmann U, Schmuttermair C, Dippel E, Koenen W, Riedel F, Sankala M, Tryggvason K, Kobzik L, Moldenhauer G, Arnold B, Goerdts S. Differential expression of a gene signature for scavenger/lectin receptors by endothelial cells and macrophages in human lymph node sinuses, the primary sites of regional metastasis. *J Pathol.* 208, 574-589, 2006
245. Nikolova, G., Jabs, N., Konstantinova, I., Domogatskaya, A., Tryggvason, K., Sorokin, L., Fässler, R., Gu, G., Gerber, H.-P., Ferrara, N., Melton, D.A., Lammert, E.: The vascular basement membrane: A niche for insulin gene expression and beta cell proliferation. *Devel. Cell*, 10, 397-405, 2006,
246. Takemoto M, He L, Norlin J, Patrakka J, Xiao Z, Petrova T, Bondjers C, Asp J, Wallgard E, Sun Y, Samuelsson T, Mostad P, Lundin S, Miura N, Sado Y, Alitalo K, Quaggin SE, Tryggvason K, Betsholtz C.: Large-scale identification of genes implicated in kidney glomerulus development and function. *EMBO J.* 25, 1160-1174, 2006
247. Chen Y, Sankala M, Ojala JR, Sun Y, Tuuttila A, Isenman DE, Tryggvason K, Pikkarainen T. A phage display screen and binding studies with acetylated LDL provide evidence for the importance of the scavenger receptor cysteine-rich (SRCR) domain in the ligand-binding function of MARCO. *J. Biol. Chem.* 281, 12767-12775, 2006
248. Mukhopadhyay S, Chen Y, Sankala M, Peiser L, Pikkarainen T, Kraal G, Tryggvason K, Gordon S. MARCO, an innate activation marker of macrophages, is a class A scavenger receptor for Neisseria meningitidis. *Eur. J. Biochem.* 69, 940-949, 2006
249. Nigatu A, Sime W, Gorfu G, Geberhiwot T, Andurén I, Ingerpuu S, Doi M, Tryggvason K, Hjendahl P, Patarroyo M. Megakaryocytic cells synthesize and platelets secrete  $\alpha$ 5-laminins, and the endothelial laminin isoform laminin 10 ( $\alpha$ 5 $\beta$ 1 $\gamma$ 1) strongly promotes adhesion but not activation of platelets. *Thromb. Haemost.* 95, 85-93, 2006.
250. Wondimu Z, Gorfu G, Kawataki T, Smirnov S, Yurchenco P, Tryggvason K, Patarroyo M. Characterization of commercial laminin preparations from human placenta in comparison to recombinant laminins 2 ( $\alpha$ 2 $\beta$ 1 $\gamma$ 1), 8 ( $\alpha$ 4 $\beta$ 1 $\gamma$ 1), 10 ( $\alpha$ 5 $\beta$ 1 $\gamma$ 1). *Matrix Biol.* 25, 89-93, 2006.
251. Fujii Y, Khoshnoodi J, Takenaka H, Hosoyamada M, Nakajo A, Bessho F, Kudo A, Takahashi S, Arimura Y, Yamada A, Nagasawa T, Ruotsalainen V, Tryggvason K, Lee AS, Yan K. The effect of dexamethasone on defective nephrin transport caused by ER stress: a potential mechanism for

- the therapeutic action of glucocorticoids in the acquired glomerular diseases. *Kidney Int.* 69, 1350-1359, 2006
252. Srivastava T, Garola RE, Kestila M, Tryggvason K, Ruotsalainen V, Sharma M, Savin VJ, Jalanko H, Warady BA. Recurrence of proteinuria following renal transplantation in congenital nephrotic syndrome of the Finnish type. *Pediatr. Nephrol.* 21, 711-718, 2006
  253. Österholm AM, He B, Pitkaniemi J, Albinsson L, Berg T, Sarti C, Tuomilehto J, Tryggvason K. Genome-wide scan for type 1 diabetic nephropathy in the Finnish population reveals suggestive linkage to a single locus on chromosome 3q. *Kidney Int.* 71, 140-145, 2007.
  254. Khoshnoodi J, Hill S, Tryggvason K, Hudson B, Friedman DB Identification of N-linked glycosylation sites in human nephrin using mass spectrometry. *J Mass Spectrom.* 4, :370-379, 2007.
  255. Patrakka J, Xiao Z, Nukui M, Takemoto M, He L, Oddsson A, Perisic L, Kaukinen A, Al-Khaliliszigyarto C, Uhlen M, Jalanko H, Betsholtz C, Tryggvason K Expression and Subcellular Distribution of Novel Glomerulus-Associated Proteins Dendrin, Ehd3, Sh2d4a, Plekhh2, and 2310066E14Rik. *J Am Soc Nephrol.* 18, 689-697, 2007.
  256. Arredouani MS, Franco F, Imrich A, Fedulov A, Lu X, Perkins D, Soininen R, Tryggvason K, Shapiro SD, Kobzik L. Scavenger receptors SR-AI/II and MARCO limit pulmonary dendritic cell migration and allergic airway inflammation. *J. Immunol.* 178, 5912-5920, 2007.
  257. Dahl M, Bauer AK, Arredouani M, Soininen R, Tryggvason K, Kleeberger SR, Kobzik L. Protection against inhaled oxidants through scavenging of oxidized lipids by macrophage receptors MARCO and SR-AI/II. *J Clin Invest.* 117, 757-764, 2007.
  258. Ojala JR, Pikkarainen T, Tuuttila A, Sandalova T, Tryggvason K. Crystal structure of the cysteine-rich domain of scavenger receptor marco reveals the presence of a basic and an acidic cluster that both contribute to ligand recognition. *J Biol Chem.* 282,1 6654-6666, 2007.
  259. He L, Sun Y, Patrakka J, Mostad P, Norlin J, Xiao Z, Andrae J, Tryggvason K, Samuelsson T, Betsholtz C, Takemoto M. Glomerulus-specific mRNA transcripts and proteins identified through kidney expressed sequence tag database analysis. *Kidney Int.* 71, 889-900, 2007
  260. Vainionpää N, Lehto VP, Tryggvason K, Virtanen I. Alpha4 chain laminins are widely expressed in renal cell carcinomas and have a de-adhesive function. *Lab Invest.* 87:780-791, 2007.
  261. Qian H, Georges-Labouesse E, Nystrom A, Domogatskaya A, Tryggvason K, Jacobsen SE, Ekblom M. Distinct roles of integrins {alpha}6 and {alpha}4 in homing of fetal liver hematopoietic stem and progenitor cells. *Blood.* 2007 Jun 22; [Epub ahead of print]
  262. Genis L, Gonzalo P, Tutor AS, Galvez BG, Martinez-Ruiz A, Zaragoza C, Lamas S, Tryggvason K, Apte SS, Arroyo AG. Functional interplay between endothelial nitric oxide synthase and membrane type 1-matrix metalloproteinase in migrating endothelial cells. *Blood,* 110, 2916-2923, 2007.
  263. Nakajo A, Khoshnoodi J, Takenaka H, Hagiwara E, Watanabe T, Kawakami H, Kurayama R, Sekine Y, Bessho F, Takahashi S, Swiatecka-Urban A, Tryggvason K, Yan K. Mizoribine Corrects Defective Nephrin Biogenesis by Restoring Intracellular Energy Balance. *J Am Soc Nephrol.* 18, 2554-2564, 2007.
  264. Kawataki T, Yamane T, Naganuma H, Rousselle P, Andurén I, Tryggvason K, Patarroyo M. Laminin isoforms and their integrin receptors in glioma cell migration and invasiveness: Evidence for a role of alpha5-laminin(s) and alpha3beta1 integrin. *Exp Cell Res.* 313, 3819-3831, 2007.
  265. Wermeling F, Chen Y, Pikkarainen T, Scheynius A, Winqvist O, Izui S, Ravetch JV, Tryggvason K, Karlsson MC. Class A scavenger receptors regulate

- tolerance against apoptotic cells, and autoantibodies against these receptors are predictive of systemic lupus. *J Exp Med.* 204, 2259-2265, 2007.
266. Tryggvason S, Nukui M, Oddsson A, Tryggvason K, Jörnvall H. Glomerulus proteome analysis with two-dimensional gel electrophoresis and mass spectrometry. *Cell Mol Life Sci.* 64:3317-3335, 2007.
267. Jakobsson L, Domogatskaya A, Tryggvason K, Edgar D, Claesson-Welsh L. Laminin deposition is dispensable for vasculogenesis but regulates blood vessel diameter independent of flow. *FASEB J.* 22, 1530-1539, 2007
268. Qiao Q, Osterholm AM, He B, Pitkäniemi J, Cordell HJ, Sarti C, Kinnunen L, Tuomilehto-Wolf E, Tryggvason K, Tuomilehto J. A genome-wide scan for type 1 diabetes susceptibility genes in nuclear families with multiple affected siblings in Finland. *BMC Genet.* 8, 84, 2007.
269. He L, Sun Y, Takemoto M, Norlin J, Tryggvason K, Samuelsson T, Betsholtz C. The glomerular transcriptome and a predicted protein-protein interaction network. *J Am Soc Nephrol.* 19:260-268, 2008.
270. Doné SC, Takemoto M, He L, Sun Y, Hultenby K, Betsholtz C, Tryggvason K. Nephric is involved in podocyte maturation but not survival during glomerular development. *Kidney Int.* 73:697-704, 2008.
271. Espada J, Varela I, Flores I, Ugalde AP, Cadiñanos J, Pendás AM, Stewart CL, Tryggvason K, Blasco MA, Freije JM, López-Otín C. Nuclear envelope defects cause stem cell dysfunction in premature-aging mice *J Cell Biol.* 181, 27-35, 2008.
272. Tulla M, Lahti M, Puranen JS, Brandt AM, Käpylä J, Domogatskaya A, Salminen TA, Tryggvason K, Johnson MS, Heino J. Effects of conformational activation of integrin alpha 1I and alpha 2I domains on selective recognition of laminin and collagen subtypes. *Exp Cell Res.* 314:1734-1743, 2008.
273. Dunér F, Patrakka J, Xiao Z, Larsson J, Vlamis-Gardikas A, Pettersson E, Tryggvason K, Hultenby K, Wernerson A. Dendrin expression in glomerulogenesis and in human minimal change nephrotic syndrome. *Nephrol Dial Transplant.* 23, 2504-2511, 2008.
274. Gorfu G, Virtanen I, Hukkanen M, Lehto VP, Rousselle P, Kenne E, Lindbom L, Kramer R, Tryggvason K, Patarroyo M. Laminin isoforms of lymph nodes and predominant role of alpha5-laminin(s) in adhesion and migration of blood lymphocytes. *J. Leukoc Biol* 84, 701-712, 2008.
275. Plantman S, Patarroyo M, Fried K, Domogatskaya A, Tryggvason K, Hammarberg H, Cullheim S. Integrin-laminin interactions controlling neurite outgrowth from adult DRG neurons in vitro. *Mol Cell Neurosci* 39, 50-62, 2008.
276. Tran-Lundmark K, Tran PK, Paulsson-Berne G, Fridén V, Soininen R, Tryggvason K, Wight TN, Kinsella MG, Borén J, Hedin U. Heparan sulfate in perlecan promotes mouse atherosclerosis: roles in lipid permeability, lipid retention, and smooth muscle cell proliferation. *Circ Res* 103, 43-52, 2008.
277. Domogatskaya A, Rodin S, Boutaud A, Tryggvason K. Laminin-511 but not -332, -111, or -411 enables mouse embryonic stem cell self-renewal in vitro. *Stem Cells* 26, 2800-2809, 2008.
278. Shimizu M, Khoshnoodi J, Akimoto Y, Kawakami H, Hirano H, Higashihara E, Hosoyamada M, Sekine Y, Kurayama R, Kurayama H, Joh K, Hirabayashi J, Kasai K, Tryggvason K, Ito N, Yan K. Expression of galectin-1, a new component of slit diaphragm, is altered in minimal change nephrotic syndrome. *Lab Invest,* 89, 178-195, 2008.
279. He B, Österholm AM, Hoverfält A, Forsblom C, Hjörleifsdóttir EE, Nilsson AS, Parkkonen M, Pitkäniemi J, Hreidarsson A, Sarti C, McKnight AJ, Maxwell AP, Tuomilehto J, Groop PH, Tryggvason K. Association of Genetic Variants at 3q22 with Nephropathy in Patients with Type 1 Diabetes Mellitus. *Am J Hum Genet* 84, 5-13. 2009.
280. Sun Y, He L, Takemoto M, Patrakka J, Pikkarainen T, Genové G, Norlin J, Truvé K, Tryggvason K, Betsholtz C. Glomerular Transcriptome Changes

- Associated with Lipopolysaccharide-Induced Proteinuria. *Am J Nephrol.* 29, 558-570, 2009.
281. Goldberg S, Harvey SJ, Cunningham J, Tryggvason K, Miner J. Glomerular filtration is normal in the absence of both agrin and perlecan-heparan sulfate from the glomerular basement membrane. *Nephrol Dial Transplant.* 24, 2044-2051, 2009.
282. Wu C, Ivars F, Anderson P, Hallmann R, Vestweber D, Nilsson P, Robenek H, Tryggvason K, Song J, Korpos E, Loser K, Beissert S, Georges-Labouesse E, Sorokin LM. Endothelial basement membrane laminin alpha5 selectively inhibits T lymphocyte extravasation into the brain. *Nat Med.* 15, 519-527, 2009.
283. Sekine Y, Nishibori Y, Akimoto Y, Kudo A, Ito N, Fukuhara D, Kurayama R, Higashihara E, Babu E, Kanai Y, Asanuma K, Nagata M, Majumdar A, Tryggvason K, Yan K. Amino acid transporter LAT3 is required for podocyte development and function. *J Am Soc Nephrol.* 20, 1586-1596, 2009.
284. Ebarasi L, He L, Hultenby K, Takemoto M, Betsholtz C, Tryggvason K, Majumdar A. A reverse genetic screen in the zebrafish identifies *crb2b* as a regulator of the glomerular filtration barrier. *Devel Biol.* 334, 1-9, 2009.
285. Bowdish DM, Sakamoto K, Kim MJ, Kroos M, Mukhopadhyay S, Leifer CA, Tryggvason K, Gordon S, Russell DG. MARCO, TLR2, and CD14 are required for macrophage cytokine responses to mycobacterial trehalose dimycolate and *Mycobacterium tuberculosis*. *PLoS Pathog.* Jun;5(6):e1000474, 2009.
286. Salo S, Boutaud A, Hansen AJ, He L, Sun Y, Morales S, Venturini A, Martin P, Nokelainen P, Betsholtz C, Mathiasen IS, Tryggvason K. Antibodies blocking adhesion and matrix binding domains of laminin-332 inhibit tumor growth and metastasis in vivo. *Int J Cancer.* 125, 1814-1825, 2009.
287. Sime W, Lunderius-Andersson C, Enoksson M, Rousselle P, Tryggvason K, Nilsson G, Harvima I, Patarroyo M. Human mast cells adhere to and migrate on epithelial and vascular basement membrane laminins LM-332 and LM-511 via alpha(3)beta(1) integrin. *J Immunol.* 183, 4657-4665, 2009.
288. Beltcheva O, Hjorleifsdottir EE, Kontusaari S, Tryggvason K. Sp1 Specifically Binds to an Evolutionarily Conserved DNA Segment within a Region Necessary for Podocyte-Specific Expression of Nephtrin. *Nephron Exp Nephrol.* 2009 Oct 9;114(1):e15-e22. [Epub ahead of print].
289. Fornoni A, Jeon J, Santos JV, Cobianchi L, Jauregui A, Inverardi L, Mandic SA, Bark C, Johnson K, McNamara G, Pileggi A, Molano RD, Reiser J, Tryggvason K, Kerjaschki D, Berggren PO, Mundel P, Ricordi C. Nephtrin is expressed on the surface of insulin vesicles and facilitates glucose stimulated insulin release. *Diabetes.* 59, 190-199, 2010.
290. Chen Y, Wermeling F, Sundqvist J, Jonsson AB, Tryggvason K, Pikkarainen T, Karlsson MC. A regulatory role for macrophage class A scavenger receptors in TLR4-mediated LPS responses. *Eur J Immunol.* 40, 1451-1460, 2010.
291. Rodin S, Domogatskaya A, Ström S, Hansson EM, Chien KR, Inzunza J, Hovatta O, Tryggvason K. Long-term self-renewal of human embryonic stem cells on human recombinant laminin-511 in xeno-free and feeder-free environment. *Nature Biotechnol.* 28, 611-615, 2010.
292. Hyvarinen J, Parikka M, Sormunen R, Ramet M, Tryggvason K, Kivirikko KI, Myllyharju J, Koivunen P. Deficiency of a transmembrane prolyl 4-hydroxylase in the zebrafish leads to basement membrane defects and compromised kidney function. *J Biol Chem.* 2010 Oct 15. [Epub ahead of print]
293. Dunér F, Lindström K, Hultenby K, Hulkko J, Patrakka J, Tryggvason K, Haraldsson B, Wernerson A, Pettersson E. Permeability, ultrastructural changes, and distribution of novel proteins in the glomerular barrier in early puromycin aminonucleoside nephrosis. *Nephron Exp Nephrol.* 2010;116(2):e42-52. Epub 2010 Jun 29.

294. Xu X, Patrakka J, Sistani L, Uhlen M, Jalanko H, Betsholtz C, Tryggvason K. Expression of Novel Podocyte-Associated Proteins *sult1b1* and *ankrd25*. *Nephron Exp Nephrol*. 2010 Aug 18;117(2):e39-e46. [Epub ahead of print]
295. Mukhopadhyay S, Varin A, Chen Y, Liu B, Trggvason K, and Gordon S. SR-A/MARCO-mediated ligand delivery enhances intracellular TLR and NLR function, but ligand scavenging from cell-surface limits TLR-4 response to pathogens. *Blood*, in press.

## II REVIEWS, BOOK CHAPTERS AND OTHER PUBLICATIONS

1. Tryggvason, K.L: Glomeruli and their basement membrane in the normal human kidney and in the congenital nephrotic syndrome of the Finnish type. Thesis: *Acta Univ. Oul.* A57, Biochem.17, 1977.
2. Liotta, L.A., Tryggvason, K., Garbisa, S., Gehron-Robey, P. and Murray, J.C.: Interactions of metastatic tumor cells with basement membrane collagen. In: *Cancer Campaign*, vol. 4, Metastatic Tumor Growth, (Grundman, E., ed.), Gustav Fischer Verlag, Stuttgart - New York, 21-30, 1980.
3. Tryggvason, K. ja Pihlajaniemi, P.: Tyvikalvojen rakenne ja aineenvaihdunta. (Review, Finnish). *Duodecim.*, 22, 1532-1542, 1980.
4. Liotta, L.A., Garbisa, S., Tryggvason, K. and Wicha, M.: Correlation of metastatic behavior with tumor cell degradation of basement mebrane collagen. In: *Tumor Progression*, 6th Chicago Cancer Symposium (Crispen, ed.), Elsevier North Holland, 49-57, 1980.
5. Tryggvason, K., Pihlajaniemi, T., Salo, T., Liotta, L.A. and Kivirikko, K. I.: Studies on the biosynthesis and degradation of type IV collagen. In: *New Trends in Basement Membrane Research* (Kühn, K., Schöne, H. and Timpl, R., eds.), Raven Press, New York, 187-193, 1982.
6. Liotta, L.A., Garbisa, S. and Tryggvason, K.: Biochemical mechanisms involved in tumor cell penetration of the basement membranes. In: *New Developments in the Study of the Biology of Metastasis* (Liotta, L.A. and Hart, I., eds.), Martin-Nijhoff Publishers, Hague - Boston - London, 319-333, 1982.
7. Tryggvason, K., Pihlajaniemi, T. and Salo, T.: Studies on the molecular composition and degradation of type IV procollagen. In: *Basement Membranes and Cell Movement*, Ciba Foundation Symposium, (Porter, R. and Whelan, J., eds.), Pitman, London, 108, pp. 117-129, 1984.
8. Myllylä, R., Pajunen, L., Höyhty, M., Turpeenniemi-Hujanen, T., Tryggvason, K. and Kivirikko, K.I.: Monoclonal antibodies to collagen hydroxylases and their use for the localization of the genes for these enzymes on human chromosomes. *Ann. N.Y. Acad. Sci.* 460, 486-488, 1985.
9. Tryggvason, K.: Pattern of basement membrane degradation by metastatic tumor cell enzymes. In: *Biochemistry and Molecular Genetics of Cancer Metastasis* (Rabson, A., Liotta, L.A. and Lapis, K., eds.), Martinus - Nijhoff, pp. 151-161, 1986.
10. Tryggvason, K., Höyhty, M. and Salo, T.: Proteolytic degradation of extracellular matrix in tumor invasion. *Biochim. Biophys. Acta, Reviews on Cancer* 907, 191-217, 1987.
11. Hostikka, S.L. and Tryggvason, K.: Comparison of the human basement membrane (type IV) collagen  $\alpha 1(IV)$  and  $\alpha 2(IV)$  chains and their genes. In: *Progress in Basement Membrane Research*. Renal and Related Aspects in Health and Disease (Ed. M.C. Gubler, M. Stemberg) John Libbey Eurotex Ltd., pp. 5-11, 1988.
12. Pikkariainen, T., Shows, T. and Tryggvason, K.: he B1 chain of human laminin. n: *Progress in Basement Membrane Research*. Renal and Related Aspects in Health and Disease (Ed. M.C. Gubler, M. Stemberg) John Libbey Eurotex Ltd., pp. 25-33, 1988.

13. Tryggvason, K.: Extracellular matrix and its enzymatic degradation in tumor invasion. (A review). In: *Influence of tumor development on the host* (L.A. Liotta, ed.) Kluwer Academic Publishers, Dordrecht, pp. 72-83, 1989.
14. Tryggvason, K., Soininen, R., Hostikka S.L., Ganguly, A., Huotari, M. and Prockop, D.J.: Structure of the human type IV collagen genes. *Ann. New York Acad. Sci.* 580, 97-111, 1990.
15. Vuolteenaho, R., Kallunki, T., Chow, L., Ikonen, J., Pikkarainen, T. and Tryggvason, K.: Genes for the human laminin B1 and B2 chains. In: *Extracellular Matrix Genes* (Eds. Boyd, C.D. and Sundell, L.) Academic Press, pp. 175-193, 1990.
16. Tryggvason, K., Huhtala, P., Tuuttila, A., Chow, L., Keski-Oja, J. and Lohi, J.: Structure and expression of type IV collagenase genes. *Cell. Diff. Devel.*, 32, 307-312, 1990.
17. Tryggvason, K.: Cloning of the Alport syndrome gene. *Ann. Med. Biol.* 23, 237-239, 1991.
18. Tryggvason, K.: Från protein till gen: Alport syndrom. *Handlingar* 135, 259-264, 1991.
19. Tryggvason, K., Huhtala, P., Höyhtyä, M., Hujanen, E. and Hurskainen, T.: 70 K type IV collagenase (gelatinase). In: *Matrix Metalloproteinases* (Ed. Birkedal-Hansen). Academic Press, pp. 45-50, 1992.
20. Huhtala, P., Chow, L., Shows, T. and Tryggvason, K.: Structure of the human 70 K type IV collagenase gene and assignment of the gene to the q21 region of chromosome 16. In: *Matrix Metalloproteinases* (Ed. Birkedal-Hansen). Academic Press, p. 84, 1992.
21. Tryggvason, K., Zhou, J., Hostikka, S.L. et Sariola, H.: Collagène de type IV dans la membrane basale glomérulaire normale et anormale. In: *Actualités néphrologiques Jean Hamburger*. (Funck-Brentano, J.L., Bach, J.F., Kreis, H. and Grünfeld, J.P. eds), Hôpital Necker, Médecine-Sciences, Flammarion, Paris, pp. 1-13, 1992.
22. Tryggvason, K., Höyhtyä, M. and Pyke, C.: Type IV collagenases in invasive tumors. *Breast Cancer Res. Treat.* 24, 209-218, 1993.
23. Tryggvason, K., Zhou, J., Hostikka, S.L. and Sariola, H.: Type IV collagen in normal and diseased glomerular basement membrane. In: *Advances in Nephrology* (Grünfeld, J.-P., Bach, J.F., Kreis, H. and Maxwell, M.H. eds.) Necker Hospital, Mosby-Year Book 22, 1-14, 1993.
24. Tryggvason, K., Zhou, J. and Hostikka, S.L.: Alport syndrome and other inherited basement membrane disorders. In: *Molecular and Cellular Aspects of Basement Membranes* (Timpl, R. and Rohrbach, D.H., eds.). Academic Press Inc., pp. 421-437, 1993.
25. Tryggvason, K.: Biochemistry and genetic diseases of glomerular basement membrane. In: *Research Opportunities and Future Directions in Nephrology* (N.A. Kurtzman, ed.). Seminars in Nephrology, 13, 447-456, 1993.
26. Tryggvason, K.: The laminin family. *Curr. Opinion. Biol.* 5, 877-882, 1993.
27. Hudson, B.G., Reeders, S.T. and Tryggvason, K.: Type IV collagen: Structure, gene organization, and role in human diseases: molecular basis of Goodpasture and Alport syndromes and diffuse leiomyomatosis. Minireview, *J. Biol. Chem.* 268, 26033-26036, 1993.
28. Burgeson, R.E., Chiquet, M., Deutzmann, R., Ekblom, P., Engel, J., Kleinman, H., Martin, G.R., Meneguzzi, G., Paulsson, M., Sanes, J., Timpl, R., Tryggvason, K., Yamada, Y. and Yurchenco, P.D.: A new nomenclature for laminins. *Matrix Biol.* 14, 209-211, 1994.
29. Hudson, B.G., Kalluri, R. and Tryggvason, K.: Pathology of glomerular basement membrane nephropathy. *Curr. Opin. Nephrol. Hypert.* 3, 334-339, 1994.
30. Munaut, C., Reponen, P., Huhtala, P., Kontusaari, S., Foidart, J.-M. and Tryggvason K.: Structure of Mouse 92-kDa type IV collagenase gene. In:

- Inhibition of Matrix Metalloproteinases: Therapeutic potential* (R.A. Greenwald, L.M. Golub, eds.) Ann. N.Y. Acad. Sci. 732, 369-371, 1994.
31. Reponen, P., Sahlberg, C., Munaut, C., Thesleff, I. and Tryggvason, K.: High expression of 92-kDa type IV collagenase (gelatinase B) in the osteoclast lineage during mouse development. In: *Inhibition of Matrix Metalloproteinases: Therapeutic potential* (R.A. Greenwald, L.M. Golub, eds.) Ann. N.Y. Acad. Sci. 732, 472-475, 1994.
  32. Thesleff, I., Sahlberg, C., Reponen, P. and Tryggvason, K.: Matrix remodeling during tooth and bone development: The expression of gelatinases A and B and TGF $\beta$  growth factors. In: *The Biological Mechanisms of Tooth Eruption, Resorption and Replacement by Implants* (D. Davidovitch, ed.). Harvard Society for the Advancement of Orthodontics, Boston, Massachusetts, 273-281, 1994.
  33. Tryggvason, K.: Molecular properties and diseases of collagens. *Kidney Int.* 47, 24-28, 1995.
  34. Tryggvason, K. and Heiskari, N.: Alport syndrome. In: *Molecular Nephrology: Kidney Function in Health and Disease* (Eds D. Schlöndorff and J. Bonventre) Marcel Dekker Inc. New York-Basel-Hong Kong, pp. 795-808, 1995.
  35. Kääriäinen, H. and Tryggvason, K.: Hereditary renal diseases and their genetic diagnosis. Editorial. *Duodecim*, 111, 1398-1400, 1995.
  36. Sariola, H., Hostikka, S.L., Lukkarila, S. and Tryggvason, K.: Distribution of type IV collagen  $\alpha$ 1,  $\alpha$ 2 and  $\alpha$ 5 chains in human tissues. In: *Molecular Genetics and Pathology of Alport Syndrome* (Ed. Karl Tryggvason), Karger, Basel, Vol. 117, pp. 130-141, 1996.
  37. Tryggvason, K.: Mutations in type IV collagen genes and Alport phenotypes. In: *Molecular Genetics and Pathology of Alport Syndrome* (Ed. Karl Tryggvason), Karger, Basel, Vol. 117, pp. 154-171, 1996.
  38. Tryggvason, K., Haakana, H., Airenne, T., Iivanainen, A. and Kallunki, T.: Laminin genes: structure, regulation and mutations in disease. In: *The Laminins* (Eds. P. Ekblom and R. Timpl), Harwood Academic Publishers, pp. 51-63, 1996.
  39. Tryggvason, K. and Ninomiya, Y.: Alport syndrome (hereditary nephritis). In: *Clinical Studies in Medical Biochemistry, 2nd edition* (Eds. Yoshifumi Ninomiya and Robert H. Glew), Oxford University Press, pp. 218-226, 1997.
  40. Tryggvason, K., Heikkilä, P., Pettersson, E., Tibell, A., Thorner, P. Can Alport syndrome be treated by gene therapy? *Kidney Int.* 51, 1493-1499, 1997.
  41. Tryggvason, K. and Heikkilä, P.: Alport Syndrome. In: *Principles of Molecular Medicine* (Ed. J. Larry Jamison), Humana Press Inc., pp. 665-668, 1998.
  42. Tryggvason, K.: Unraveling the mechanisms of glomerular ultrafiltration: Nephritin, a key component of the slit diaphragm. *J. Am. Soc. Nephrol.* 10, 2440-2445, 1999.
  43. Tryggvason, K., Ruotsalainen, V. and Wartiovaara, J.: Discovery of the congenital nephrotic syndrome gene discloses the structure of the mysterious molecular sieve of the kidney. *Int. Devel. Biol.* 43, 445-51, 1999.
  44. Holmberg, C., Jalanko, H., Tryggvason, K. and Rapola, J.: Congenital nephrotic syndrome. In: *Pediatric Nephrology*, 4<sup>th</sup> ed, edited by Barratt TM, Avner ED, Harmon WE, Baltimore, Lippincott Williams & Wilkins, pp 765-777, 1999.
  45. Heikkilä, P., Tryggvason, K., and Thorner, P.: Animal models of Alport syndrome: Advancing the prospects for effective human gene therapy. Minireview. *Exp. Nephrol.* 8, 1-7, 2000.
  46. Kraal, G., van der Laan, L.J., Elomaa, O. and Tryggvason, K.: The macrophage receptor MARCO. *Microbes Inf.* 2, 313-316, 2000.
  47. Tryggvason, K., and Martin, P.: Alport syndrome and basement membrane collagen. In: *The Metabolic and Molecular Basis of Inherited Disease*. 8<sup>th</sup> ed. Edited by Schriver, CR, Beaudet, AL, Sly, WS, Valle, D, Childs, B. Kinzler, KW. and Vogelstein B., McGraw-Hill, pp. 5453-5466, 2001.

48. Khoshnoodi, J. and Tryggvason, K.: Congenital nephrotic syndromes. *Curr. Opin. Genet. Devel.* 11, 322-327, 2001.
49. Tryggvason, K. and Wartiovaara, J.: Molecular basis of glomerular permselectivity. *Curr. Opin. Nephrol. Hypertension*, 10, 543-549, 2001.
50. Jalanko, H., Holmberg, C. and Tryggvason, K.: Diseases of the glomerular filtration barrier: Alport syndrome and congenital nephrosis (NPHS1). In: *The kidney – from normal development to congenital disorders*. (eds. P. Vize, J. Baard, A. Woolf), Academic Press, London, pp. 475-492, 2001.
51. Jalanko, H., Patrakka, J., Tryggvason, K., and Holmberg, C.: Genetic kidney diseases disclose the pathogenesis of proteinuria. *Ann. Med.* 33, 526-533, 2001.
52. Tryggvason, K.: Nephrin: role in normal kidney and in disease. *Advances in Nephrology*, Mosby, Inc. vol 31, 2001.
53. Tryggvason, K.: Nephrin – role in normal kidney and in disease. *Adv. Nephrol.*, Necker Hospital, pp. 221-234, 2001
54. Khoshnoodi, J., and Tryggvason, K.: Unraveling the molecular make-up of the glomerular podocyte slit diaphragm. *Exp. Nephrol.* 9, 355-359, 2001.
55. Patarroyo, M., Tryggvason, K., and Virtanen, I.: Laminin isoforms in tumor invasion, angiogenesis and metastasis. *Semin. Cancer Biol.* 12, 197-207, 2002.
56. Patarroyo M, Tryggvason K, Virtanen I.: Laminin isoforms in tumor invasion, angiogenesis and metastasis. *Semin Cancer Biol*, 12, 197-207, 2002.
57. Tryggvason, K.: Primary hereditary nephropathies. In: *The Genetics of Renal Disease*, (eds. F. flinter, E. Maher, A. Saggarr-Malik), Oxford University Press, Oxford, pp.167-182, 2003.
58. Hudson, B.G., Tryggvason, K., Sundaramoorthy, M., and Neilson, E.G.: Alport's syndrome, Goodpasture's syndrome, and type IV collagen. *N Engl J Med.* 348, 2543-2556, 2003.
59. Tryggvason, K. and Pettersson, E.: Causes and consequences of proteinuria: the kidney filtration barrier and progressive renal failure. *J Intern Med.* 254, 216-224, 2003.
60. Holmberg, C., Tryggvason, K., Kestilä, M. and Jalanko, H.: Congenital nephrotic syndrome. In: *Pediatric Nephrology* (eds. E. Avner, W Harmon, P Niaudet) Lippincott Williams & Wilkins, pp. 503-516, 2003.
61. Tryggvason, K.: Primary hereditary nephropathy. In: *The genetics of renal disease* (eds F Flinter, E Maher, A Saggarr-Malik) Oxford University Press, pp 167182, 2003.
62. Tryggvason, K., Wartiovaara, J. How does the kidney filter plasma? *Physiology* 20:96-101, 2005.
63. Aumailley, M., Bruckner-Tuderman, L., Carter, W.G., Deutzmann, R., Edgar, D., Ekblom, P., Engel, J., Engvall, E., Hohenester, E., Jones, J.C., Kleinman, H.K., Marinkovich, M.P., Martin, G.R., Mayer, U., Meneguzzi, G., Miner, J.H., Miyazaki, K., Patarroyo, M., Paulsson, M., Quaranta, V., Sanes, J.R., Sasaki, T., Sekiguchi, K., Sorokin, L.M., Talts, J.F., Tryggvason, K., Uitto, J., Virtanen, I., von der Mark, K., Wewer, U.M., Yamada, Y., Yurchenco, P.D.: A simplified laminin nomenclature. *Matrix Biol.* 24, 326-332, 2005.
64. Tryggvason, K., Patrakka, J.: Thin basement membrane nephropathy. *J Am Soc Nephrol.* 17, 813-822, 2006.
65. Tryggvason, K., Patrakka, J., and Wartiovaara, J.: Hereditary proteinuria syndromes and mechanisms of proteinuria. *N. Engl. J. Med.* 354, 1387-1401, 2006.
66. Tryggvason K, Pikkarainen T, Patrakka J. Nck links nephrin to actin in kidney podocytes. *Cell* 125, 221-224, 2006.
67. Patrakka, J. and Tryggvason, K. Nephrin - Role in Renal Physiology and Disease. Review. *Trends Mol. Med.* 13, 396-403, 2007.
68. Betsholtz C, He L, Takemoto M, Norlin J, Sun Y, Patrakka J, Tryggvason K. The glomerular transcriptome and proteome. *Nephron Exp Nephrol.* 106, 32-36, 2007.



69. Gross O, Borza DB, Anders HJ, Licht C, Weber M, Segerer S, Torra R, Gubler MC, Heidet L, Harvey S, Cosgrove D, Lees G, Kashtan C, Gregory M, Savige J, Ding J, Thorner P, Abrahamson DR, Antignac C, Tryggvason K, Hudson B, Miner JH. Stem cell therapy for Alport syndrome: the hope beyond the hype. *Nephrol Dial Transplant*. 24, 731-734, 2009.
70. Patrakka J, Tryggvason K. New insights into the role of podocytes in proteinuria. *Nature Rev. Nephrology*, 5, 463-468, 2009

### III PATENTS

1. Tryggvason, K. and Liotta, L.A.: Detecting malignant cells with monoclonal antibodies specific to type IV collagenase enzyme. US patent no. 4,677,058, June 30, 1987.
2. Tryggvason, K and Liotta, L.A.: Immunological method for the determination of metastatic cell activity. Canadian patent no 1,230,052, August 12, 1987.
3. Tryggvason, K. and Liotta, L.A.: Antibody composition for the detection of malignant cells with metastatic cell activity. US patent no. 4,808,528, February 28, 1989.
4. Tryggvason, K. and Liotta, L.A.: Basement membrane collagen degrading enzyme and method of purifying same. US patent no. 4,816,400, March 28, 1989.
5. Tryggvason, K. and Hostikka, S.L.: A method for determining the nucleotide sequence of a novel human type IV collagen  $\alpha 5(\text{IV})$  chain gene. US patent no. 5,114,840, May 19, 1992.
6. Tryggvason, K., Hostikka, S.L. and Höyhty, M.: Immunohistological methods for detection of the human type IV collagen  $\alpha 5$  chain. US patent no. 5,354,690, October 11, 1994.
7. Tryggvason, K., and Liotta, L.A.: Immunological methods for the detection of the human type IV collagen  $\alpha 5$  chain. US patent no 5,593,900, January 14, 1997.
8. Tryggvason, K., Kallunki, P. and Pyke, C.: Laminin  $\gamma$ -2: Diagnostic and therapeutic use. US patent no. 5,660,982, August 26, 1997.
9. Tryggvason, K., Elomaa, O. and Kangas, M.: An isolated DNA sequence for a novel macrophage receptor with a collagenous domain and the polypeptide chain encoded by such a DNA sequence. US patent no. 5,691,197, November 25, 1997.
10. Tryggvason, K., Kallunki, P. and Pyke, C.: Laminin chains: diagnostic and therapeutic use. Australian patent no 699,183, October 13, 1998.
11. Tryggvason, K., Lukkarinen, O., Heikkilä, P. and Parpala, T.: Perfusion apparatus and methods for pharmaceutical delivery. US patent no. 5,871,464, February 16, 1999.
12. Tryggvason, K., Kallunki, P. and Pyke, C.: Laminin chains: diagnostic and therapeutic use. European patent no 784,703, July 14, 1999.
13. Tryggvason, K., Kallunki, P. and Pyke, C.: Laminin chains: diagnostic and therapeutic use. US patent no 6,143,505, November 7, 2000.
14. Tryggvason, K., Elomaa, O. and Kangas, M.: Macrophage receptor with a collagenous domain US patent no. 6,063,901, May 16, 2000
15. Tryggvason, K., Kestilä, M., Lenkkeri, U., and Männikkö, M.: Nephron gene and protein. US patent no 6,207,811, March 27, 2001
16. Tryggvason, K., Lukkarinen, O., Heikkilä, P. and Parpala, T.: Method for viral vector delivery US patent no. 6,342,214, January 29, 2002.
17. Tryggvason, K., Hostikka, S.L. and Zhou, J.: Method for determining the nucleotide sequence of the gene for the  $\alpha 5(\text{IV})$  chain of the human type IV collagen. US patent no 6,576,418, June 10, 2003

18. Tryggvason, K., Lukkarinen, O., Heikkilä, P. and Parpala, T. Perfusion apparatus and methods for pharmaceutical delivery US patent no. 6,638,264, October 28, 2003
19. Tryggvason, K., Kestilä, M., Lenkkeri, U., and Männikkö, M.: Nephtrin gene and protein. Australian patent pending.
20. Tryggvason, K., Kestilä, M., Lenkkeri, U., and Männikkö, M.: Nephtrin gene and protein. Canadian patent pending.
21. Tryggvason, K., Kestilä, M., Lenkkeri, U., and Männikkö, M.: Nephtrin gene and protein. European patent pending.
22. Tryggvason, K., Kestilä, M., Lenkkeri, U., and Männikkö, M.: Nephtrin gene and protein. Japanese patent pending.
23. Tryggvason, K., Kestilä, M., Lenkkeri, U., and Männikkö, M.: Nephtrin gene and protein. Mexican patent pending.
24. Tryggvason, K., Kestilä, M., Lenkkeri, U., and Männikkö, M.: Nephtrin protein localization to the glomerular filtration barrier. International patent pending.
25. Tryggvason, K., Kallunki, P. and Pyke, C.: Laminin chains: diagnostic and therapeutic use. Canadian patent pending.
26. Korttesmaa, J., and Tryggvason, K. Laminin 8 and methods for its use. United States Patent no 6,638,907, October 28, 2003
27. Tryggvason, K., Lukkarinen, O., Heikkilä, P. and Parpala, T.: Perfusion apparatus and methods for pharmaceutical delivery. US patent no. 6,689,090 B1, February 10, 2004.
28. Tryggvason, K., Doi, M., and Thyboll, J. Isolated laminin 10. US Patent no 6,933,273, August 23, 2005.
29. Tryggvason, K., Kallunki, K., and Pyke, C. Laminin chains: diagnostic uses. United States Patent no 6,955,924, October 18, 2005
30. Tryggvason, K., Kestilä, M., Lenkkeri, U., and Mannikko, M. Nephtrin gene and protein. United States Patent no 7,105,291, September 12, 2006
31. Tryggvason, K., Kallunki, P. and Pyke, C.: Method for inhibiting the migration of cancer cells. US patent pending.
32. Tryggvason, K., Domogatskaya, A., and Rodin, S.: Composition and method for enabling expansion and maintenance of pluripotent stem cells. U.S. Patent pending.
33. Tryggvason, K., and Salo, S.: Antibodies against domains of laminin-332 inhibit cancer cell migration and metastasis. PCT application, US Patent pending.