Invited Lecture

2015. 5. 15. (FRI) 17:10 ~ 18:00

Future Perspective of Matrix Biology and Connective Tissue Research

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Chair: Prof. Kazuki Nabeshima
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1965. Liceo Classico Garibaldi, Maturita' Classica, 1965 (M.A.)
1969. Università degli Studi di Palermo, 1969 (D.Sc.)
1972. Research Associate, Department of Human Genetics and Development-
      Dr. Arthur Bank laboratory, Columbia University, New York, NY
1979. Associate Professor, Department Obstetrics and Gynecology,
      UMDNJ-Rutgers Medical School, Piscataway, NJ
1986. Professor, Department of Microbiology and Immunology
      SUNY, Health Science Center at Brooklyn, New York, NY
1989. Dr. Amy and James Elster Professor of Molecular Biology (Connective Tissue
       Diseases), Mount Sinai School of Medicine, New York, NY
1999. Dean for Research, Mount Sinai School of Medicine, New York, NY
2002. Chief Scientific Officer and St. Giles Chair of Genetics, Hospital for Special
       Surgery, New York, NY
2005. Laura Gallagher Endowed Professor and Director, Child Health Institute of
       New Jersey, UMDNJ-Robert W. Johnson Medical School, Piscataway, NJ
2008. Dr. Amy and James Elster Professor (Connective Tissues Diseases),
       Departments of Pharmacology and System Therapeutics, and Medicine,
       Mount Sinai School of Medicine, New York, NY
1996. Co-organizer of the Keystone Symposium "Molecular and Developmental Biology of the Extracellular Matrix".
2004. President, American Society Matrix Biology
2013. Vice President, President-elect, International Society for Matrix Biology
2015. President, International Society for Matrix Biology

**SUMMARY**

In contrast to "cellular diseases", such as HIV, in which one or few cell types are sufficient to drive the whole system to a pathological state, "tissue diseases" like fibrotic conditions are driven by altered interactions between the cells and the extracellular matrix (ECM). Marfan syndrome (MFS) is a prototypical tissue disease caused by mutations in a structural component of the ECM (fibrillin-1) that are associated with secondary cellular and molecular abnormalities. Indeed, studies of mouse models of MFS have demonstrated the multiple roles of fibrillin-1 assemblies (microfibrils and elastic fibers) in supporting organ function and homeostasis. To be specific, characterization of cardiovascular and musculoskeletal manifestations in MFS mice has revealed the following previously unsuspected functions:

1. Fibrillin-1 is a component of the ECM/sarcomere multi-protein complex that regulates cardiac muscle adaptation to demanding loading conditions by modulating the AT1R/β-arrestin 2/Erk1/2 mechanosignaling cascade.
2. Fibrillin-1 is responsible for the biological and physical properties that control postnatal growth and adult homeostasis of the ascending aorta.
3. Fibrillin-1 specifies how tendon tissue transmits skeletal muscle pull to regulate bone linear growth.
4. Fibrillin-1 regulates MSC fate by modulating TGFβ bioavailability within the structural microenvironment of bone marrow niches.

As we achieve a deeper understanding of the reciprocal interactions between cells and ECM, we will be in a better position to develop new evidence-based therapies against the progression of age-related diseases of tissue degeneration.