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Abstract

Mutations in fibrillin-1 cause Marfan syndrome (MFS), the most common heritable disorder of connective tissue. Fibrillin-1 assemblies (microfibrils and elastic fibers) represent a unique dual-function component of the architectural matrix. The first role is structural for they endow tissues with tensile strength and elasticity, transmit forces across them and demarcate functionally discrete areas within them. The second role is instructive in that these macroaggregates