

# Fibrous Proteins

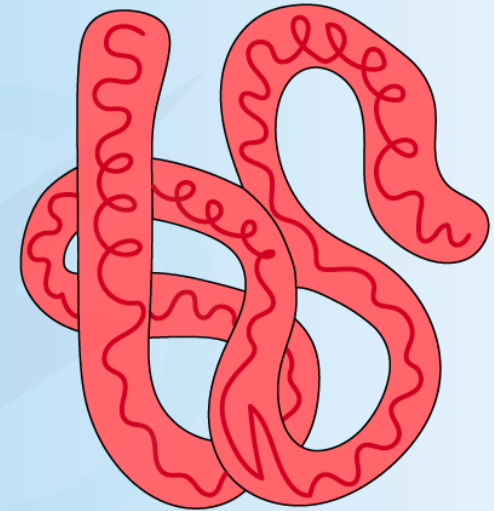
Tess Warchalowski

# Basics

- Polypeptide chains organized along an axis to produce long fibers / sheets
- **Structural support to cells**
- Characterized by highly repetitive aa sequences
- Fibrous proteins= general term
  - ie keratin, collagen, myosin, elastin



FIBROUS

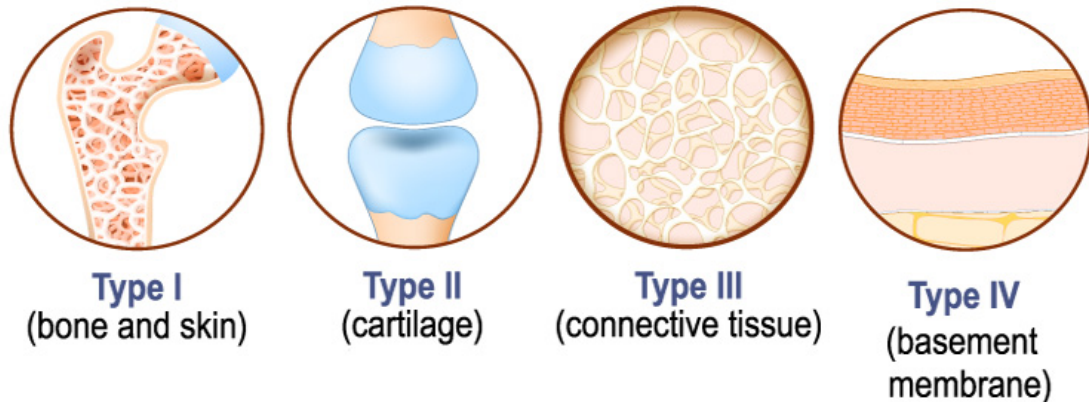
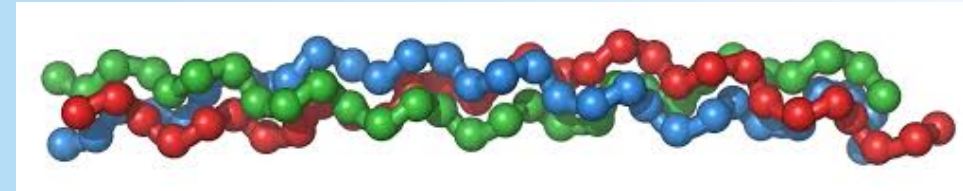


GLOBULAR

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# Collagen

- Structural protein
- Collagen= glycine, proline, lysine
- Repeating **GLY-X-Y**
- Three long alpha chains= “triple helix”
- Produced by fibroblasts ( also chondrocytes and osteoblasts)



## *TYPES*

Be So Totally Cool, Read Books

I: Bone, Skin, Tendon

II: Cartilage

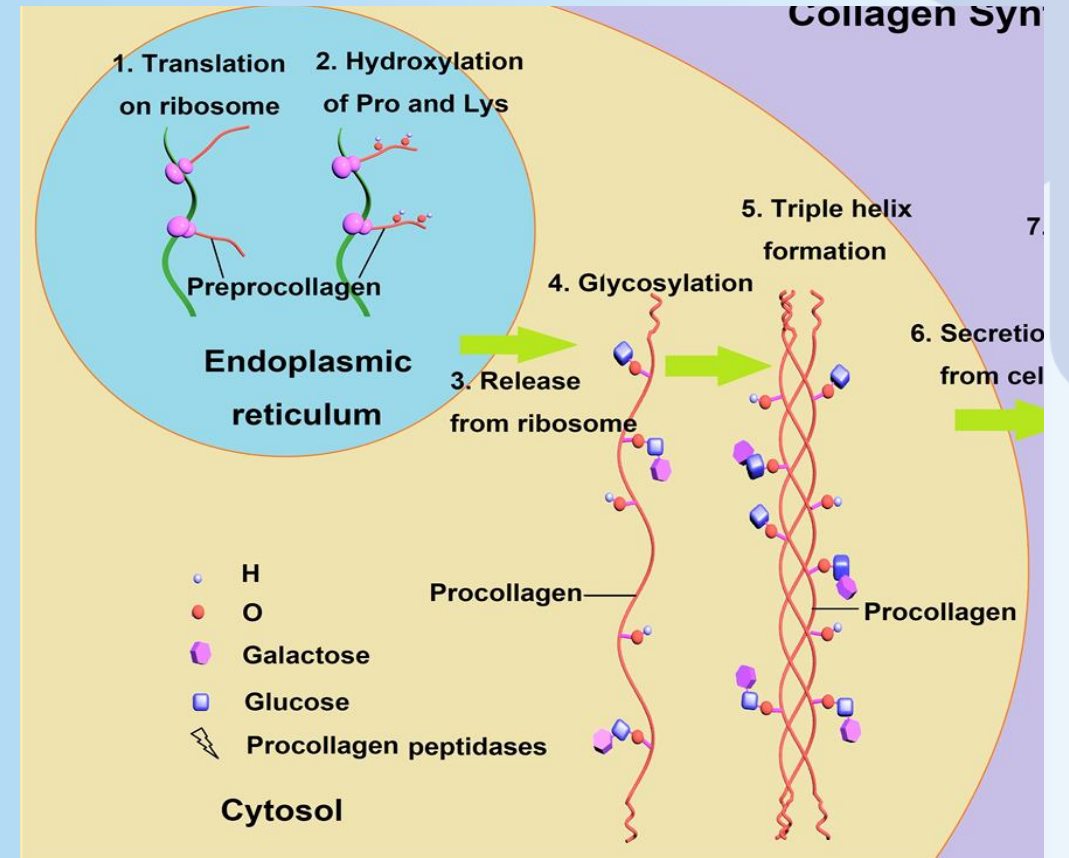
III: Reticulin and Blood vessels

IV: Basement membrane



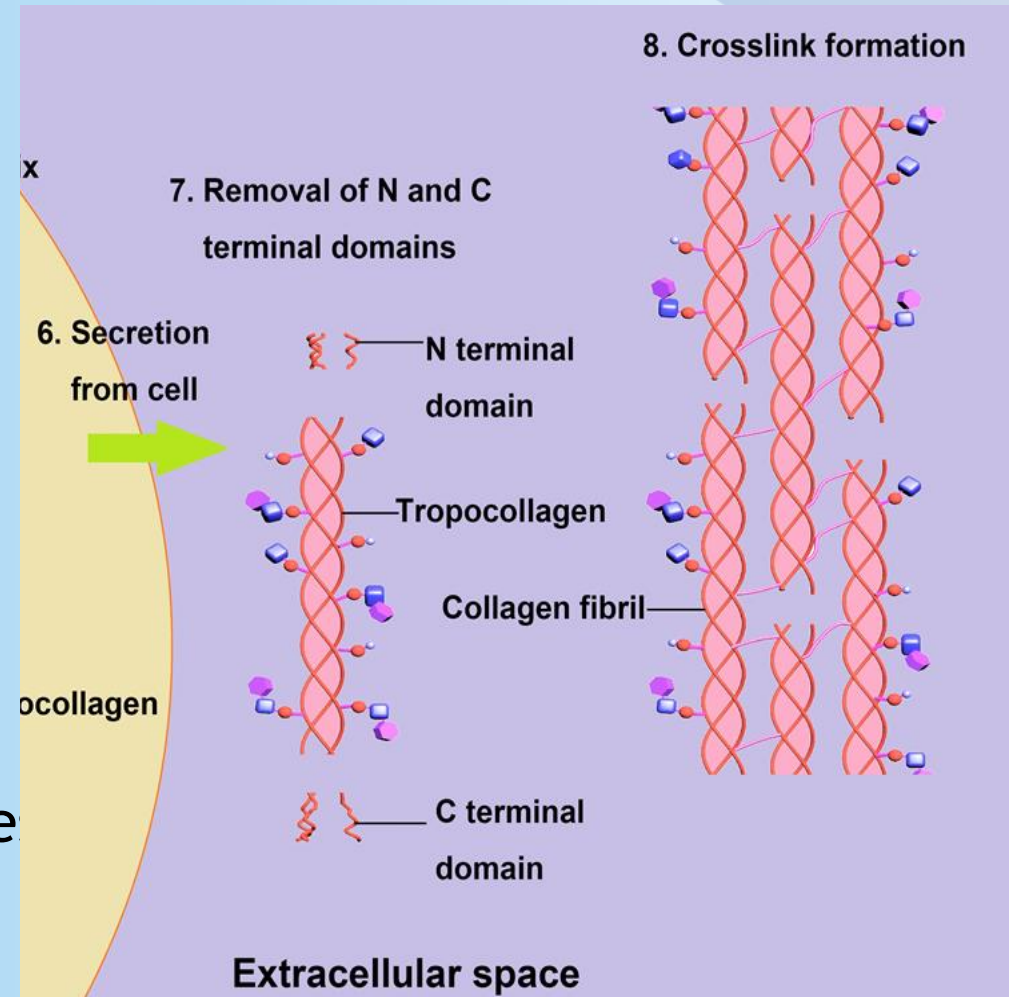
# Collagen Synthesis

1. Pro alpha chain formation in RER ( pre pro collagen)
2. Hydroxylation of proline and lysine  
- Requires vitamin C
3. Glycosylation of some hydroxylysine residues
4. 3 pro alpha chains combine= procollagen= triple helix

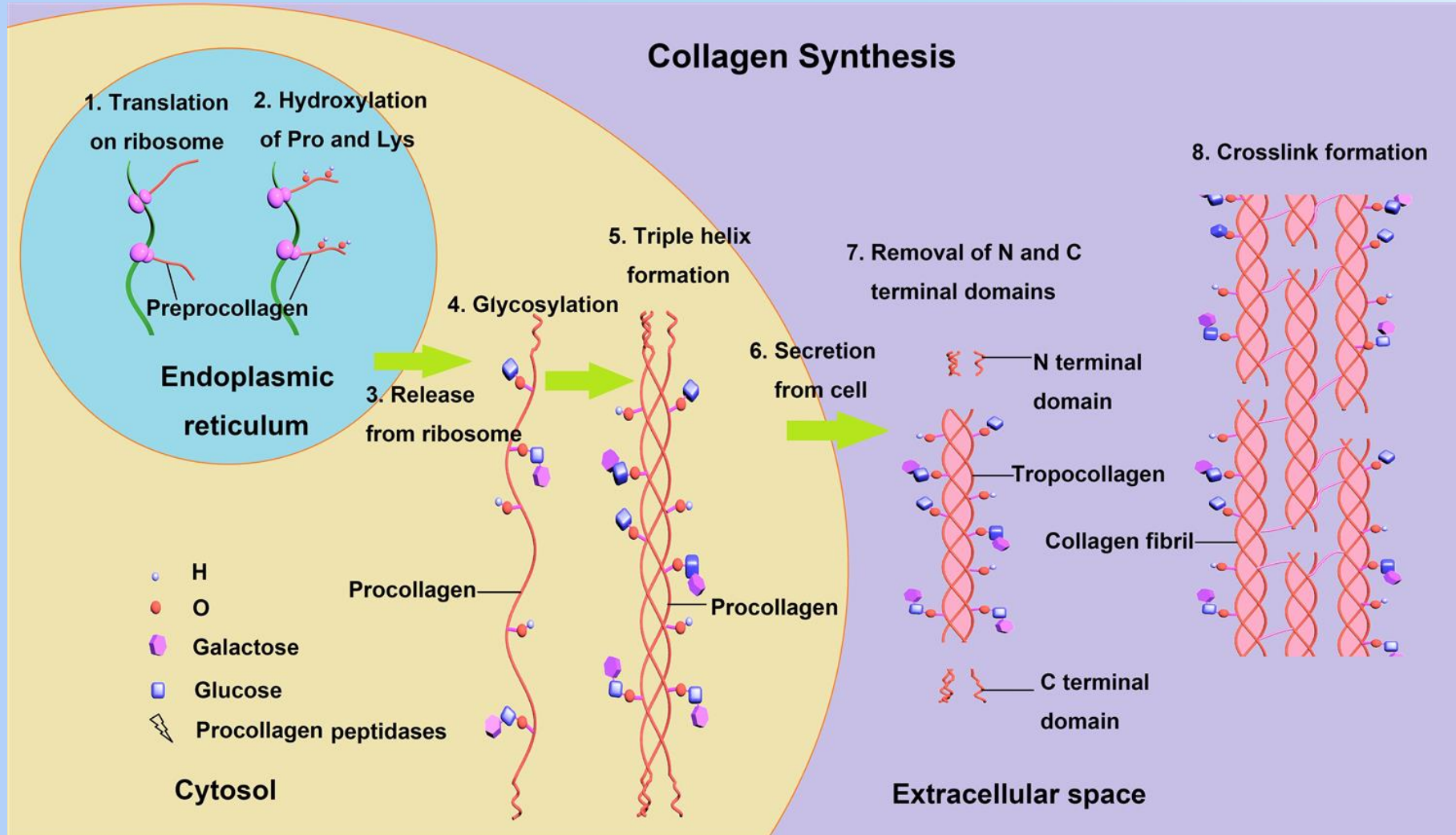


# Collagen Synthesis

5. Procollagen transported out of cells
6. Pro peptides ( N and C terminals) cleaved=  
Tropocollagen
  - problems with cleavage= Ehlers Danlos syndrome
7. Tropocollagen fibrils cross link
  - Helped by **lysyl oxidase**
  - Copper is a cofactor of lysyl oxidase
8. Collagen fibers are formed! - bundle of triple helices



# Collagen Synthesis



# Scurvy

= **Vitamin C deficiency!**

- hydroxylation inhibited
- Defective formation of collagen triple helix
- Symptoms
  - Fragile blood vessels
  - Bleeding gums !
  - Decreased immune response
  - Corkscrew hair



# Osteogenesis imperfecta

- Bones fracture easily- often with no identifiable cause
- **Mutation in genes encoding collagen chains**
- Multiple subtypes ranging in severity

## SYMPTOMS

### BITE

Bones- fractures

I (eye)- blue sclera

Teeth- dental changes

Ear- hearing loss



# Ehlers Danlos Syndrome

- Group of inheritable disorders, caused by faulty collagen synthesis
- Symptoms
  - Hypermobility of joints
  - Tendency to bleed (bruising)
  - Hyperextensible skin



## TYPES

### Hypermobility ( joint)

- most common type

### Classic type ( joint and skin)

- mutation in type V collagen synthesis

### Vascular type

- Deficient in type III procollagen

# Elastin

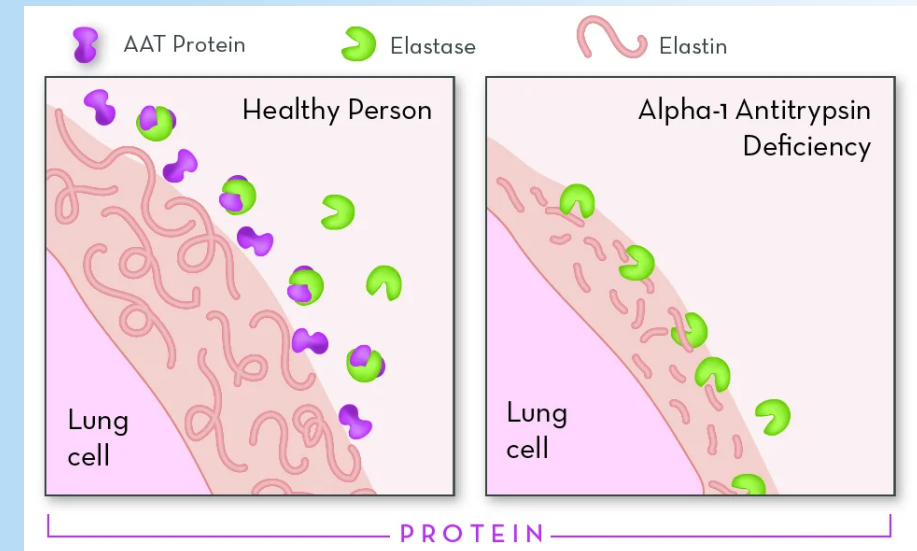
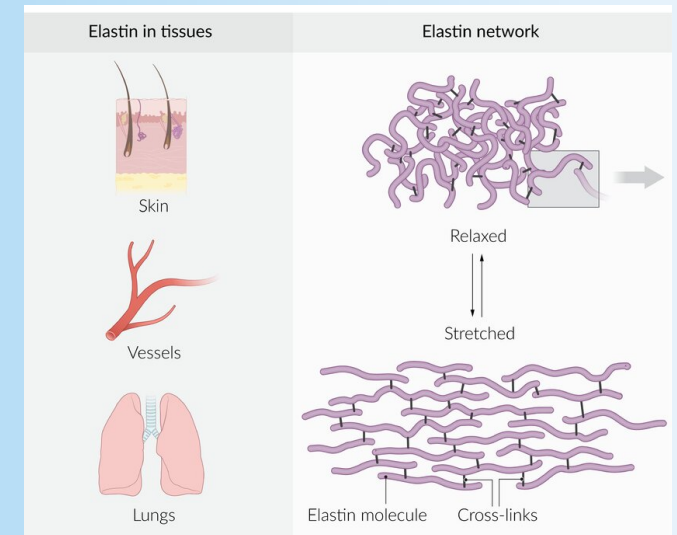
## Primary structure

- Rich in non-hydroxylated glycine, proline and lysine residues

## Alpha-1 antitrypsin deficiency

Elastase= Breaks down elastin

- **Alpha 1 AT inhibits elastase**
- Development of pulmonary emphysema
- Elastase levels are increased in smoking, inflammation, infections



## Quiz time :)



1

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