Fibrous Proteins

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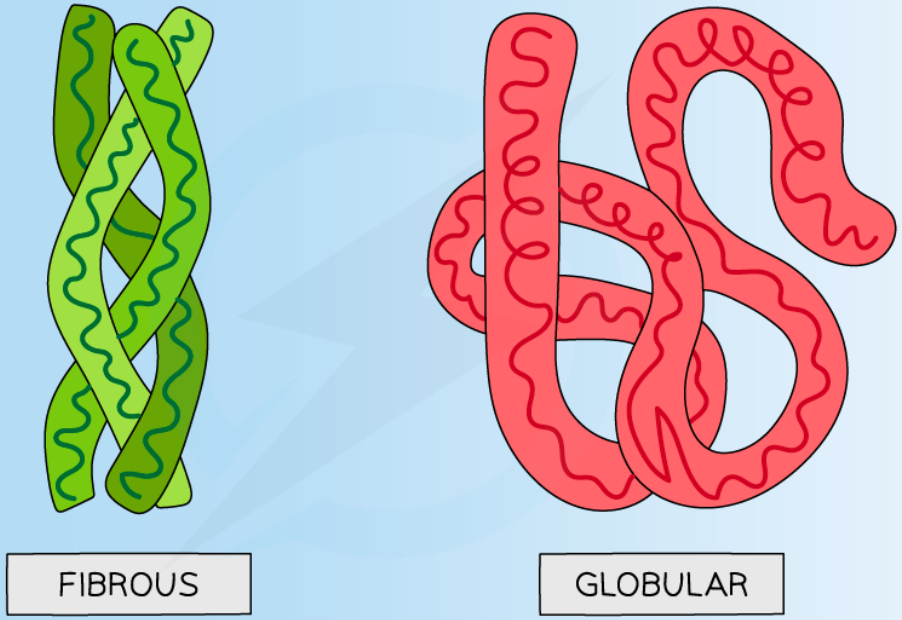
Basics

- 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



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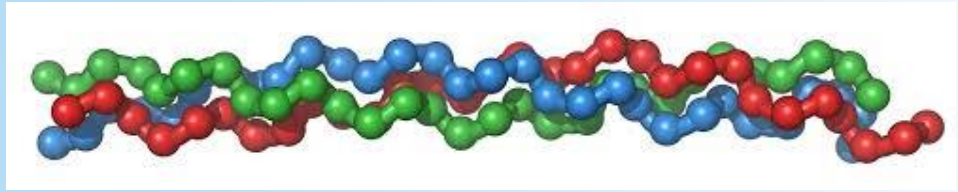
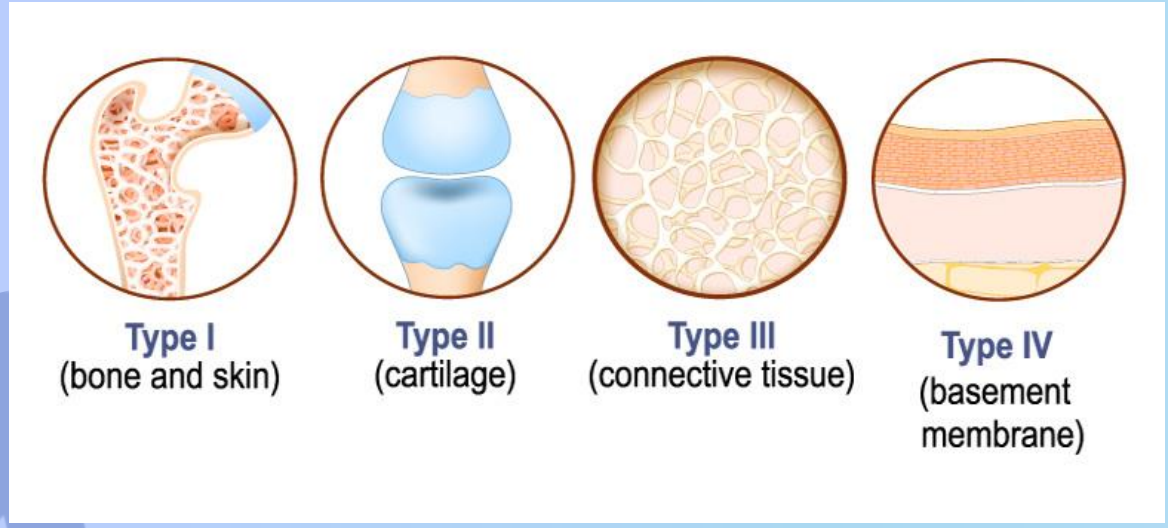
Collagen

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





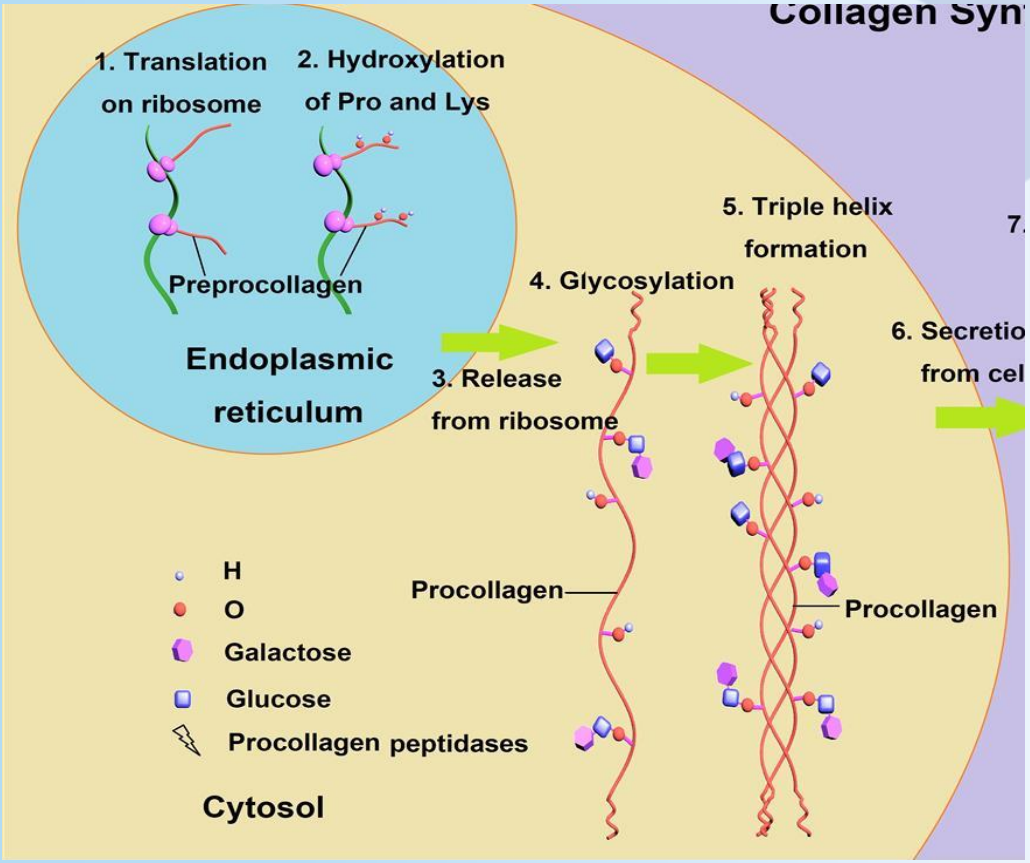
TYPES

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

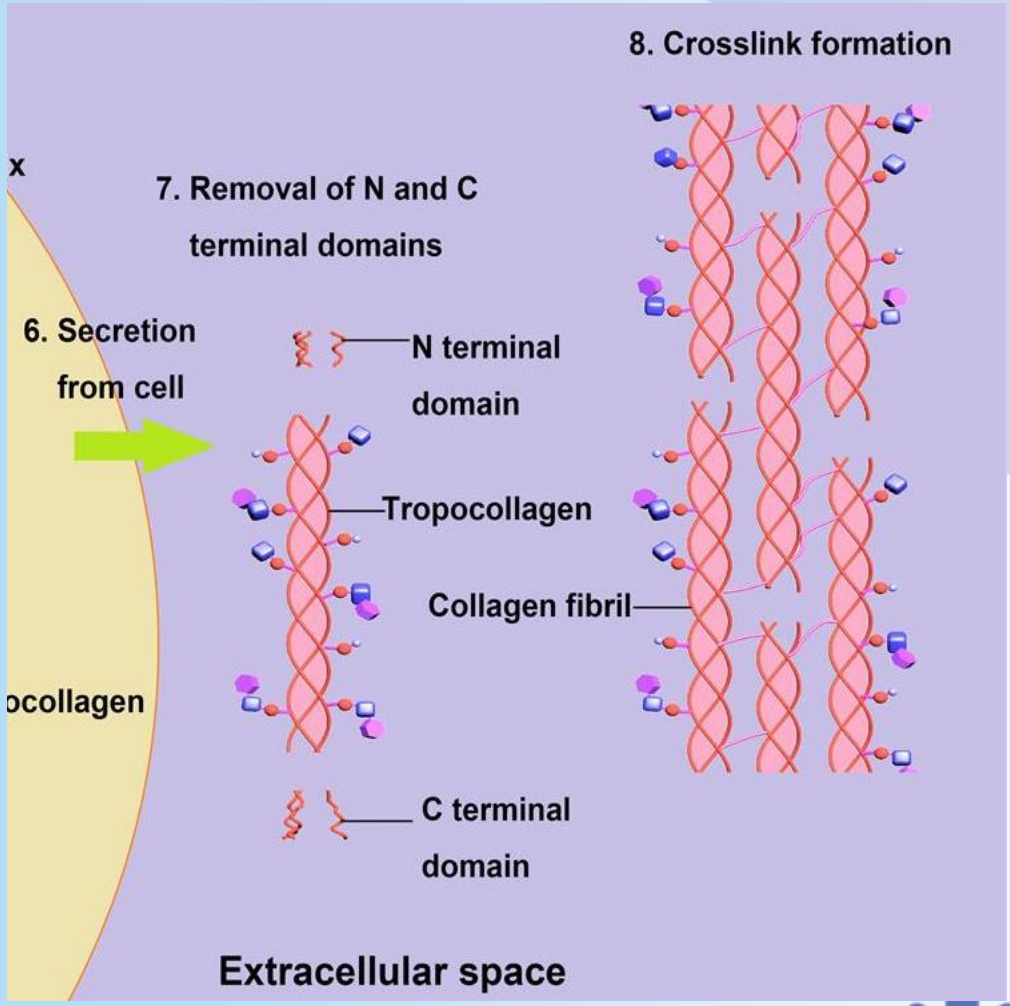
- problems with cleavage= Ehlers Danlos syndrome

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



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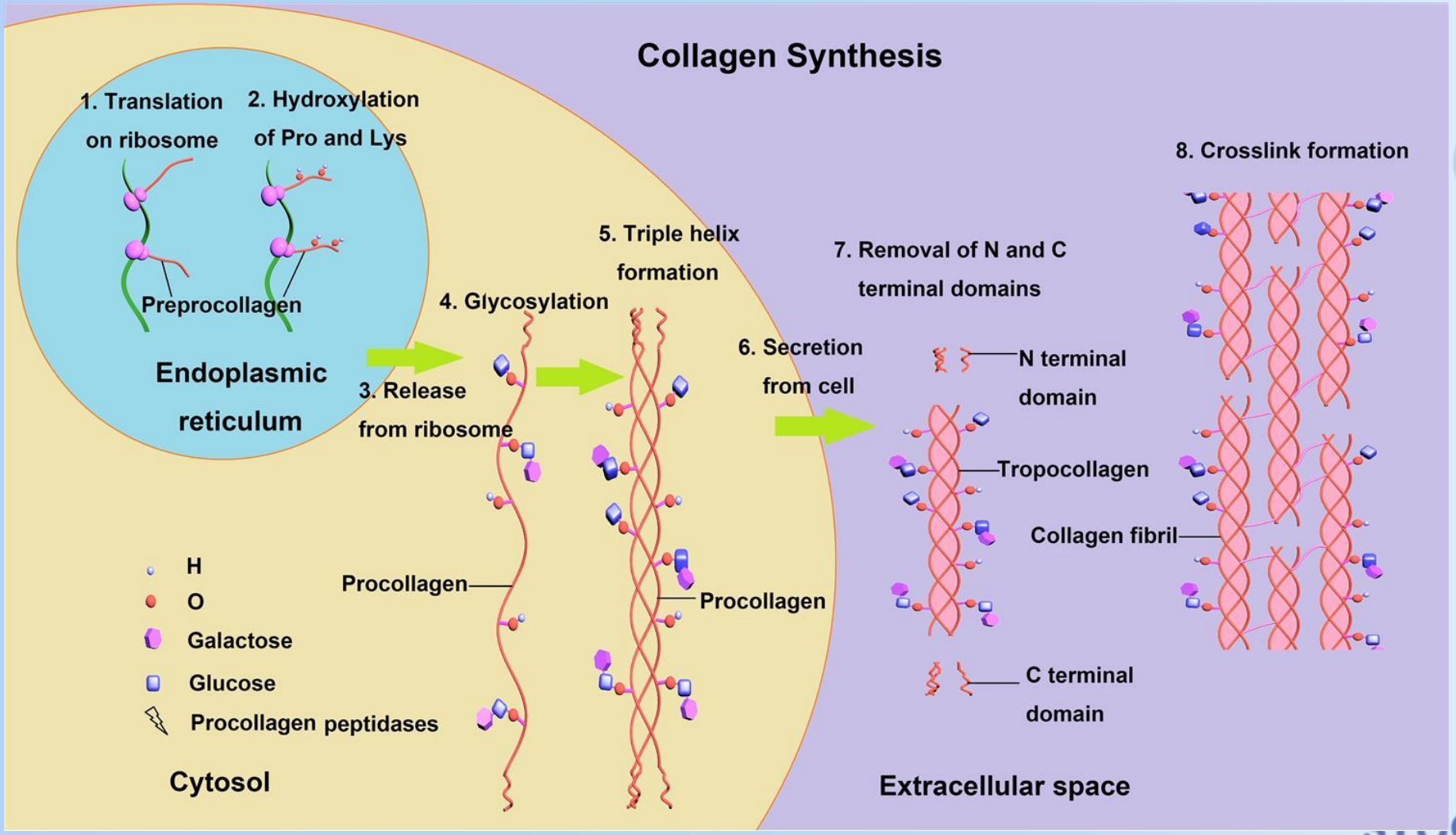
Collagen Synthesis



Scurvy

= Vitamin C deficiency!

- **proline hydroxylation inhibited**

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

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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

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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

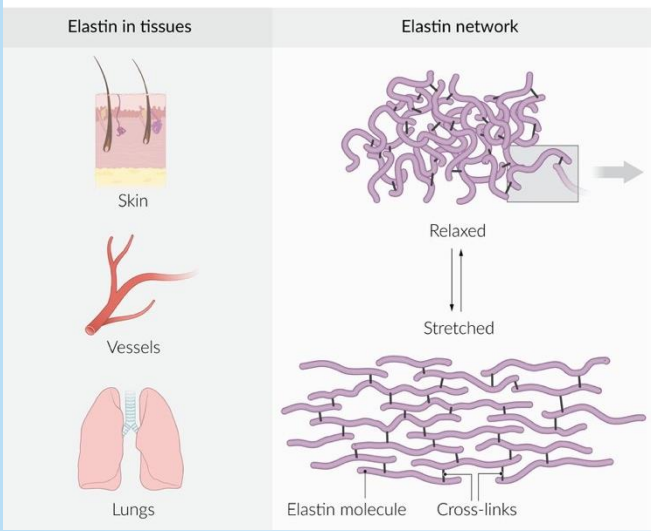
Classic type (joint and skin)
- mutation in type V collagen synthesis

Vascular type
- Deficient in type III procollagen

Elastin

Primary structure

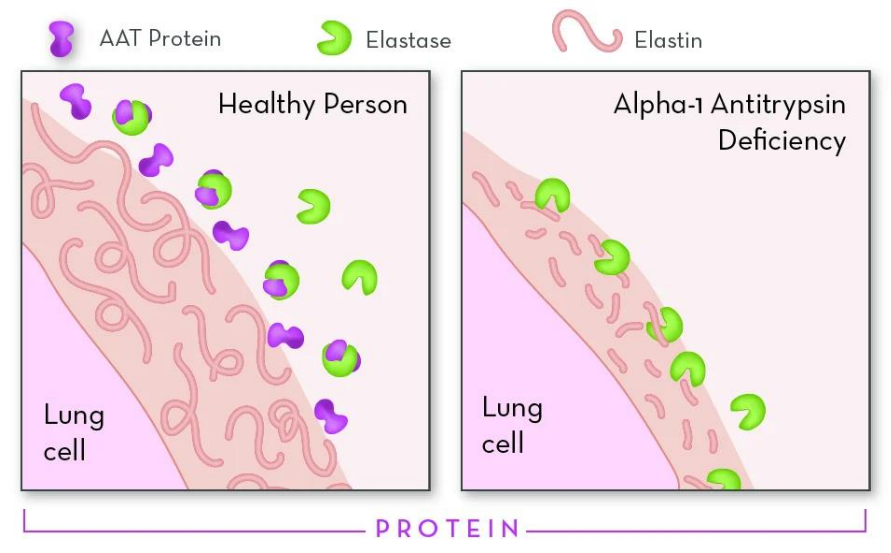
- Rich in non- hydroxylated glycine, proline and lysine residues



Alpha- 1 antitrypsin deficiency

Elastase= Breaks down elastin

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



Quiz time :)

1 Go to wooclap.com

2 Enter the event code in the top banner

Event code
PSGTBD

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