matrix goo

8 marks.

→ seen in mucous secretion.

Mucopolysaccharides or GAGs.

- previously called as mucopolysaccharides because they were isolated from mucous secretions
- principal structural component of ground substance.
- Structural element of tissues like bone, cartilage, connective tissue, cornea and synovial fluid.
- mucopolysaccharides in combination with proteins form mucoproteins.

Other functions.

- Structural Components - Collagen and elastin.

- Due to large numbers of charged groups (sulphate, carboxyl, acetylated amino groups)

↓

polysaccharide chains are kept apart.

↓

Hence can hold large amount of water & occupying space.

↓

which attribute the cushioning and lubricating effect.

eg: Hyaluronic acid in Synovial fluid.

GAGs contains: aminosugars & uronic acid.

Classification.

Sulphated

- Keratan Sulphate
- Dermatan sulphate.
- Heparan Sulfate.
- Chondroitin Sulfate.

Non Sulphated.

Hyaluronic acid.

Hyaluronic acid.

- Present in connective tissues, tendons, synovial fluid Vitreous humor.
- Serve as lubricants in joint cavities.
- repeating units → Glucuronic acid & N-acetyl glucosamine.

Chondroitin Sulfate. (CS)

- In cartilage, tendon, bone etc.
- rep. units : glucuronic acid.
  N-acetyl galactosamine Sulphate.

Keratan Sulphate. (KS)

- In tendons and Cornea.
- ★ Only GAGs which doesn't contain Uronic acid residue. (1 mark)
- rep units: Galactose
  N-acetyl glucosamine.

Heparin.

- Intravascular anticoagulant
- highly sulphated.
- rep. units: Iduronic acid
  Sulphated glucosamine.
- produced by mast cells of liver and stored in granules.
- Found in thymus, spleen, walls of large arteries and small amounts in blood.

Heparan Sulfate. (HS)

- present in aorta & amyloid tissue.
- Same str as Heparin.

Dermatan Sulfate. (DS)

- in skin, blood vessels, heart valves.
- contain: Iduronic acid and N-acetyl galactosamine.

Clinical Significance.

~~Mucopolysaccharidoses.~~ Mucopolysaccharidoses.

- ~~disease~~
- Included in group of lysosomal storage disorder.
- characterized by accumulation of mucopolysaccharide in Organs due to deficiency of lysosomal enzyme. which degrade GAGs.
- This results in Enlargement of Organs, disturbance in bone structure, skin etc.
- Mucopolysaccharides are excreted in large amount in Urine.

Disease	Defective enzyme.	GAG in Urine.
Hurler's	L Iduronidase	DS, HS
Hunter's	Iduronate Sulfatase	DS, HS.
Sanfilippo's	N-acetyl glucosaminidase	HS.
Morquio's	Galactosamine Sulfatase	KS, CS.
Maroteaux	NAc Gal 4 Sulfatase	DS.
Sly's	Beta glucuronidase.	DS, HS.

Clinical features.

- Mental retardation, growth deficiency and skeletal dysplasia are common features
- Inherited as autosomal recessive except hunter's disease, which is X linked.