



*Prix  
coup de cœur  
du Public !*

2<sup>ème</sup> Journée Scientifique de l'IRMIT  
07 décembre 2020



*Prix  
coup de cœur  
du Public !*

## Serum Bikunin: a biomarker of inborn errors of proteoglycan biosynthesis and Golgi homeostasis defects

**Walid Haouari**

Arnaud Bruneel, François Foulquier, Valérie Cormier-Daire, Christian Poüs

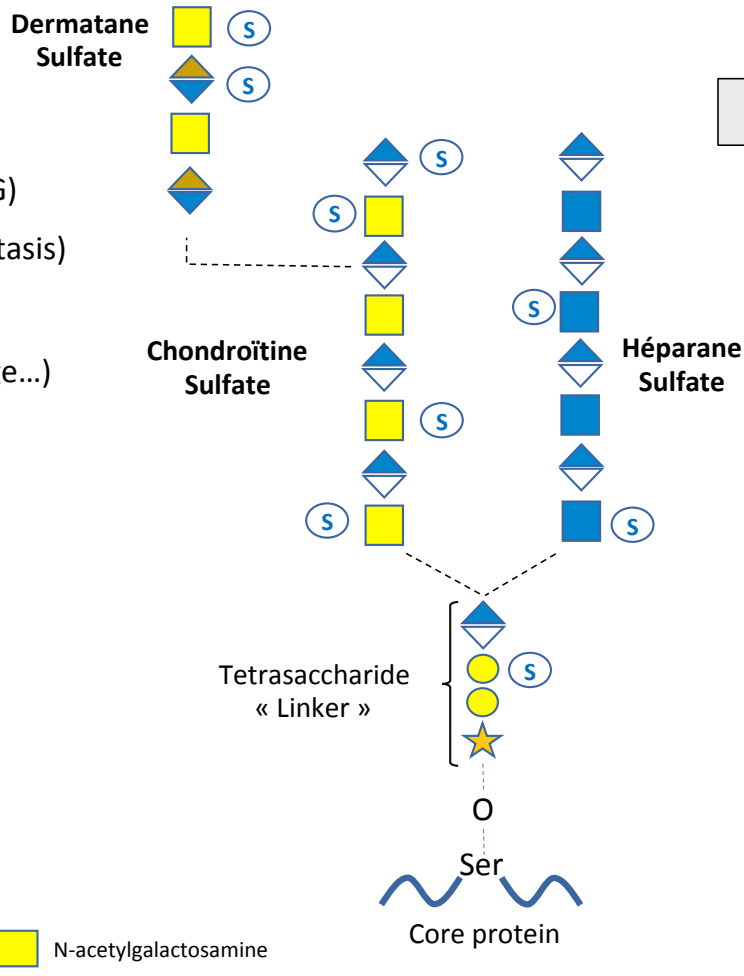
INSERM UMR1193, Université Paris-Sud, Faculté de Pharmacie Châtenay Malabry

CNRS UMR 8576 – UGSF, Université de Lille

INSERM 1163 Institut Imagine, Paris, France

# Proteoglycans

- PG = core protein + glycosaminoglycan (GAG)
- Biosynthesis in the Golgi (enzymes, homeostasis)
- Hydrophilic et viscoelastic properties of extracellular matrices (bones, joints, cartilage...)
- Immune response and cell proliferation



## Linkeropathies



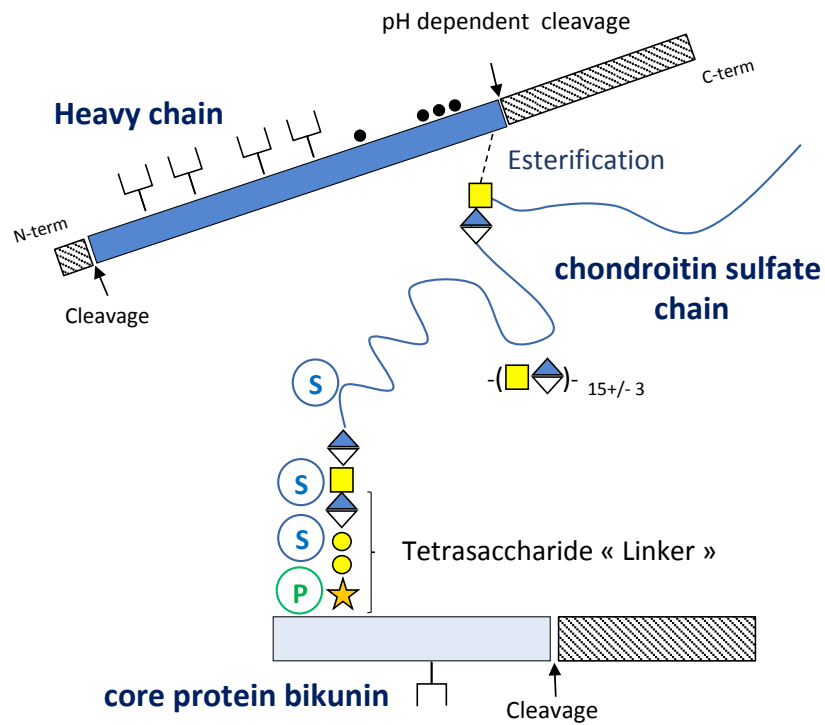
Osteo-articular symptoms

Other unspecific symptoms

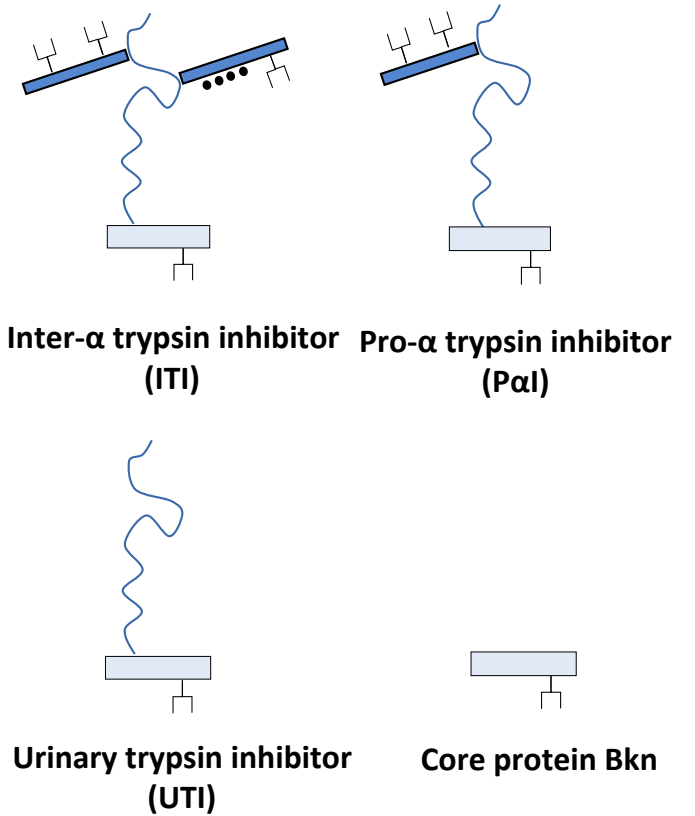
Difficult screening

# Bikunin:

A new serum biomarker in the screening of linkeropathies and Golgi homeostasis defects

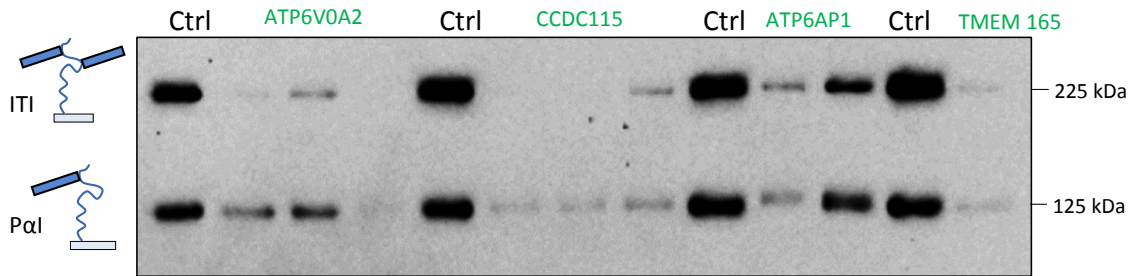
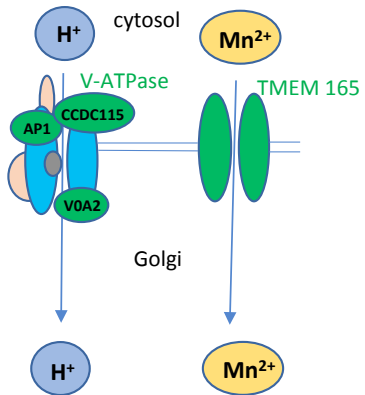
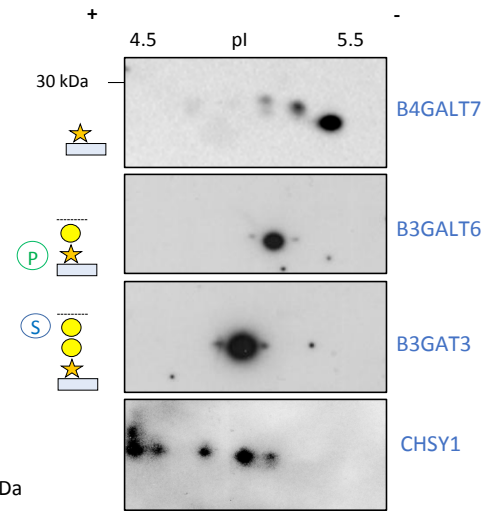
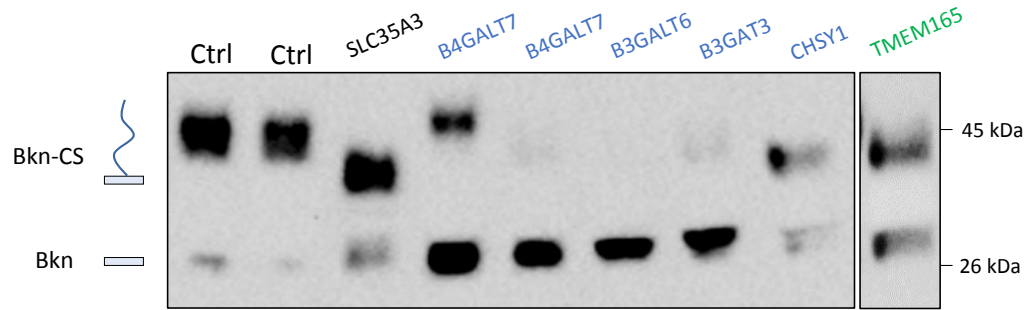
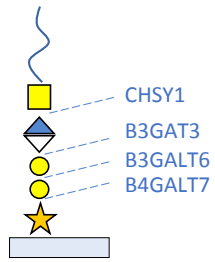


Golgi post-translational modifications



Serum isoforms

# Bikunin analysis

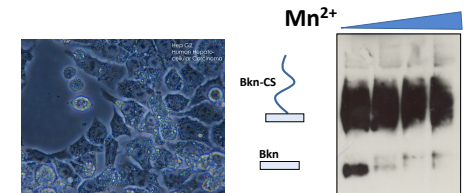


Signature profiles

Versatile biomarker

Proteoglycan biosynthesis defects

Golgi homeostasis



# Acknowledgments

## Project coordinators

Dr. Arnaud Bruneel  
Pr. Valérie Cormier-Daire  
Pr. Christian Poüs



## INSERM U1193

*“Mécanismes cellulaires et moléculaires de l’adaptation au stress et cancérogénèse”*

Charles Roseau  
Pierre Prada  
Rizk Bennani  
Sara Muhamad  
Alexandre Raynor

Samra Ouaras  
Isabelle Cantaloube  
Ameetha Ratier  
Christian Poüs

## Collaborators

Pr. François Foulquier



## UFR Pharmacie Chatenay-Malabry

