Hycult Biotech
Let's improve health

Complement dysregulation is a key driver in rare kidney diseases, where uncontrolled activation of the alternative pathway can lead to tissue damage. A central player in this process is **C3 nephritic factor (C3NeF)**, an autoantibody that stabilizes the C3 convertase and thereby prolongs complement activation. This mechanism is particularly relevant in diseases such as C3 glomerulopathy (C3G), atypical hemolytic uremic syndrome (aHUS) and membranoproliferative glomerulonephritis (MPGN), where excessive complement activity can result in severe renal damage.

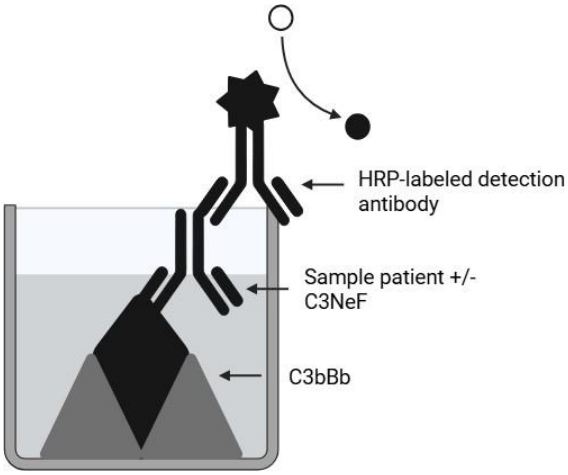
To support research in this field, Hycult Biotech offers new tools to study both C3NeF presence and C3 convertase levels.

If your research focuses on complement activation and disease mechanisms, this information is for you.

BioConcept
Paradiesrain 14
CH-4123 Allschwil 1
Postfach
Switzerland
Tel. +41 (0)61 486 80 80
Fax +41 (0)61 486 80 00
info@bioconcept.ch
www.bioconcept.ch

Content:

- Our newest C3NeF detection ELISA kit for screening C3NeF autoantibodies
- C3NeF and its role in alternative pathway dysregulation
- The importance of the C3bBb(P) convertase in complement activation
- C3bBbP ELISA kit for the determination of alternative pathway convertase levels
- Interesting publications about C3NeF and rare kidney diseases



C3NeF detection, Human, ELISA kit

The C3NeF detection ELISA kit (Cat. No. **HK3051**) is designed for the qualitative detection of C3NeF autoantibodies and supports research into alternative pathway dysregulation and complement-mediated diseases.

The C3NeF detection ELISA kit is in the final stages of development and will be available soon.

C3NeF (C3 Nephritic Factor)

C3NeF is an autoantibody that targets and stabilizes the C3 convertase (C3bBb) of the alternative complement pathway, leading to continuous complement activation and increased consumption of C3. Persistent dysregulation of this pathway is associated with rare complement-mediated kidney diseases such as C3 glomerulopathy (C3G), membranoproliferative glomerulonephritis (MPGN), and atypical hemolytic uremic syndrome (aHUS). These disorders are characterized by chronic inflammation and complement deposition in the kidneys, which may ultimately impair kidney function.

Early detection in rare kidney diseases

C3NeF-associated diseases, such as C3 glomerulopathy, often develop in children and young adults, sometimes after a period of good health. Early symptoms like blood or protein in the urine, swelling, or reduced kidney function can be mild, which may delay diagnosis.

Disease progression varies widely: some patients remain stable for years, while others experience gradual kidney function loss. This reflects underlying dysregulation of the complement system and highlights the need for better disease understanding.

As a result, there is growing interest in targeted therapies that act on the complement system, particularly the alternative pathway. Ongoing clinical studies are investigating inhibitors of key components such as C3 and factor B to reduce complement overactivation and protect kidney function.

Because early signs can be subtle and patients are often young, early recognition is crucial.

The role of C3bBb and C3bBb(P) in C3NeF-mediated complement activation

C3NeF stabilizes the alternative pathway C3 convertase (C3bBb), including its properdin-bound form, leading to continuous complement activation and excessive C3 cleavage.

Normally, this enzyme is short-lived and tightly regulated, but in C3NeF-positive patients it becomes abnormally stabilized, causing persistent immune activation.

This mechanism is central to diseases such as C3 glomerulopathy and is an important target for research and future therapies aimed at restoring normal complement regulation and protecting kidney function.

C3bBbP, Human, ELISA kit

Elevated or dysregulated C3bBbP activity is associated with complement-mediated disorders such as C3G and aHUS. As a functional readout of alternative pathway activity, C3bBbP may serve as a biomarker for complement dysregulation and support the development of targeted therapeutic strategies.

The human C3bBbP ELISA kit (Cat. No. **HK3002**) is to be used for the in vitro quantitative determination of C3bBbP in serum and plasma samples.

Interesting publications about C3NeF and C3G

“Challenges in diagnostic testing of nephritic factors”

This review highlights the complexity of detecting nephritic factors (NeFs), autoantibodies that stabilize complement convertases and contribute to complement-mediated kidney diseases such as C3 glomerulopathy. The paper discusses the different types of NeFs, current detection methods, and the need for assay standardization to improve diagnostics, patient stratification, and the development of complement-targeted therapies. The setup and principle of our assay are also discussed in this review paper.

Read here https://pubmed.ncbi.nlm.nih.gov/36451820/

“Overactivity of alternative pathway convertases in patients with complement-mediated renal diseases”

This study shows that C3NeF stabilizes the alternative pathway C3 convertase, leading to persistent complement activation and contributing to diseases such as C3 glomerulopathy and aHUS. It is particularly relevant as it links C3NeF directly to disease mechanisms and potential patient stratification and targeted therapies.

Read here https://pubmed.ncbi.nlm.nih.gov/29670616/

“Long-term follow-up including extensive complement analysis of a pediatric C3 glomerulopathy cohort'“

This study shows that C3NeF is commonly found in children with C3 glomerulopathy and is linked to ongoing complement dysregulation and variable disease progression. It highlights the value of complement profiling for understanding disease severity and guiding future targeted therapies.

Read here https://pubmed.ncbi.nlm.nih.gov/34476601/

“Different aspects of classical pathway overactivation in patients with C3 glomerulopathy and immune complex-mediated membranoproliferative glomerulonephritis”

This study shows that C3NeF stabilizes the alternative pathway C3 convertase, leading to persistent complement activation and low C3 levels, and is frequently associated with C3 glomerulopathy. It highlights the heterogeneity of C3NeF and its relevance for understanding disease mechanisms and complement-mediated kidney injury.

Read here https://pubmed.ncbi.nlm.nih.gov/34456924/