

Hydroxylysine

Table of Contents

summary

Chemical Structure

Sources

- Dietary Sources

- Biological Role

Clinical Significance

- Ehlers-Danlos Syndrome

- Hydroxylysine and Bone Health

- Therapeutic Implications

Research and Studies

- Future Directions

- Overview of Hydroxylysine Research

- Enzymatic Function and Metabolism

- Clinical Implications

Check <https://storm.genie.stanford.edu/article/1162232> for more details

Stanford University Open Virtual Assistant Lab

The generated report can make mistakes.

Please consider checking important information.

The generated content does not represent the developer's viewpoint.

summary

Hydroxylysine is a naturally occurring amino acid derivative of lysine, characterized by the addition of a hydroxyl (-OH) group at the fifth position of the lysine backbone. Its molecular formula is $C_6H_{14}N_2O_3$, with a molecular weight of approximately 162.19 g/mol, and it predominantly exists as the (5R) stereoisomer found in collagen, a critical protein for maintaining the structural integrity of connective tissues in the human body.^{[1][2]} Hydroxylysine is formed through the enzymatic action of lysyl hydroxylase, which is dependent on vitamin C, emphasizing the importance of proper nutrition for collagen synthesis and overall tissue health.^{[3][4]}

The role of hydroxylysine in collagen stability is vital; it facilitates cross-linking between collagen fibers, thereby contributing to the tensile strength and resilience of various connective tissues.^{[5][4]} Abnormal hydroxylysine levels have been asso-

ciated with several connective tissue disorders, including Ehlers-Danlos syndrome, where insufficient hydroxylation leads to weakened collagen structure and increased tissue fragility.[\[6\]\[7\]](#) Additionally, hydroxylysine has been identified as a potential biomarker for bone health, with its urinary excretion levels correlating with conditions such as osteoporosis.[\[8\]\[9\]](#)

Controversies surrounding hydroxylysine primarily involve its clinical implications and therapeutic potential. Research has indicated that dietary intake of lysine can influence hydroxylysine availability and subsequently affect collagen health, yet the full extent of its impact on connective tissue disorders remains an area of ongoing investigation.[\[10\]\[11\]](#) Furthermore, there is growing interest in the potential benefits of hydroxylysine supplementation for enhancing collagen synthesis and aiding recovery from injuries, although more research is needed to establish standardized therapeutic applications.[\[12\]\[13\]](#)

In summary, hydroxylysine plays a crucial role in collagen metabolism and structural integrity, making it a significant focus of study in relation to connective tissue health and associated disorders. As research continues to unravel its biochemical pathways and clinical relevance, hydroxylysine may offer new insights into therapeutic strategies for enhancing collagen function and treating related diseases.[\[14\]\[15\]\[16\]](#)

Chemical Structure

Hydroxylysine (Hyl) is an amino acid with the molecular formula $C_6H_{14}N_2O_3$, which includes a hydroxyl (-OH) group substituted at the 5th position of the lysine backbone, distinguishing it from its precursor lysine[\[1\]\[2\]](#). It possesses a molecular weight of approximately 162.19 g/mol, and the stereochemistry of hydroxylysine is primarily characterized by its (5R) stereoisomer, commonly found in collagen[\[2\]\[3\]](#). The chemical structure of hydroxylysine can be described by the linear formula $NH_2CH_2CH(OH)CH_2CH_2CH(NH_2)COOH \cdot HCl$, which illustrates its configuration as an amino acid[\[17\]\[2\]](#).

The structural variation of hydroxylysine includes its formation from lysine through enzymatic hydroxylation by lysyl hydroxylase enzymes, which facilitate the introduction of the hydroxyl group[\[2\]\[3\]](#). This hydroxyl modification contributes to the amino acid's functional properties, particularly its role in stabilizing collagen fibers within connective tissues due to increased hydrogen bonding capabilities[\[3\]](#). Additionally, hydroxylysine's additional oxygen atom in its composition accounts for its higher polarity compared to standard lysine derivatives[\[5\]\[1\]](#).

Sources

Hydroxylysine is a derivative of the amino acid lysine that plays significant roles in collagen structure and stability. This compound is predominantly found in collagen-rich tissues, which are essential for maintaining the integrity of connective tissues in the body[\[5\]](#).

Dietary Sources

Although hydroxylysine itself is not directly found in food sources, its precursor, lysine, is abundant in high-protein foods. Foods rich in lysine include eggs, red meat, poultry, soy products, beans, peas, and certain fish like cod and sardines[\[10\]](#). These dietary sources provide the necessary lysine that can be hydroxylated to produce hydroxylysine during collagen synthesis in the body.

Biological Role

In the context of collagen synthesis, hydroxylysine contributes to the stability of collagen fibers through cross-linking, which is crucial for the tensile strength of tissues[\[4\]](#). The post-translational modification of lysine to hydroxylysine is facilitated by the enzyme lysyl hydroxylase, which requires vitamin C as a cofactor, highlighting the importance of nutrition in the biosynthesis of this amino acid derivative[\[4\]](#). Thus, adequate dietary intake of lysine-rich foods is essential for maintaining collagen health and overall structural integrity in various tissues.

Clinical Significance

Hydroxylysine, a post-translationally modified amino acid, plays a crucial role in the structure and stability of collagen, which is vital for the integrity of connective tissues in the body. Abnormal levels of hydroxylysine have been implicated in various disorders, particularly those affecting connective tissue metabolism, such as Ehlers-Danlos syndrome (EDS) and hydroxylysine-deficient collagen disease[\[6\]\[7\]](#).

Ehlers-Danlos Syndrome

Ehlers-Danlos syndrome is a group of heritable connective tissue disorders characterized by skin hyperextensibility, joint hypermobility, and tissue fragility. In type VI of this syndrome, a deficiency of lysyl hydroxylase results in decreased hydroxylysine levels, leading to structural weaknesses in collagen[\[18\]\[19\]](#). Patients often present with symptoms such as congenital hypotonia, lax joints, and friable skin[\[20\]\[7\]](#). The identification of biochemical abnormalities associated with these conditions aids in their diagnosis and management[\[21\]\[6\]](#).

Hydroxylysine and Bone Health

In addition to its role in connective tissue, hydroxylysine levels are also significant in bone health. Research indicates that measuring hydroxylysine can serve as a biomarker for assessing bone disorders, including osteoporosis. The total urinary excretion of hydroxylysine is elevated in certain bone diseases, which correlates with the degradation of collagen in bone tissue[\[8\]\[2\]](#). As osteoporosis continues to be a major global health issue, understanding the role of hydroxylysine in bone metabolism could enhance early detection and treatment strategies[\[9\]\[22\]](#).

Therapeutic Implications

The clinical implications of hydroxylysine extend to potential therapeutic avenues. Supplements containing hydroxyproline, another collagen component, have been suggested to support overall connective tissue health and may aid in recovery from injuries or surgeries[\[12\]\[13\]](#). By enhancing collagen synthesis and stability, these therapies could provide significant benefits for patients suffering from conditions related to collagen insufficiency, highlighting the importance of hydroxylysine in clinical practice.

Research and Studies

Future Directions

Ongoing research is focused on elucidating the precise biochemical pathways involving hydroxylysine, particularly in the context of metabolic disorders. As our understanding of hydroxylysine's role in health continues to evolve, future studies may yield novel insights into therapeutic strategies aimed at ameliorating the effects of its deficiency and optimizing collagen function in various clinical scenarios[\[5\]\[14\]](#).

Overview of Hydroxylysine Research

Hydroxylysine, a derivative of lysine, has garnered attention in various fields of research due to its role in collagen stability and metabolism. This amino acid is generated from L-lysine through enzymatic biotransformation and plays a critical role in the post-translational modifications of collagen, impacting its structural integrity and biological functions[\[15\]\[23\]](#).

Enzymatic Function and Metabolism

Research has indicated that deficiencies in hydroxylysine can arise from decreased enzymatic hydroxylation of lysyl residues, affecting collagen synthesis and stability. This has implications for understanding certain connective tissue disorders and potential therapeutic approaches for diseases related to collagen metabolism[\[16\]\[24\]](#). Studies highlight the significance of enzymes involved in the hydroxylation process, as they are crucial for maintaining the structural properties of collagen, which in turn affects tissue integrity and repair mechanisms[\[14\]\[25\]](#).

Clinical Implications

The clinical relevance of hydroxylysine is evident in conditions associated with collagen defects, where hydroxylysine metabolism is disrupted. For instance, disorders related to lysine and hydroxylysine metabolism are categorized under a group of rare diseases that necessitate further investigation into their underlying mechanisms and potential treatments[\[11\]](#). Research into hydroxylysine has also explored its dietary impacts, suggesting that adequate levels of lysine can influence hydroxylysine availability and subsequently affect collagen health[\[10\]](#).

References

- [1]: [Showing metabocard for 5-Hydroxylysine \(HMDB0000450\)](#)
- [2]: [Bone biomarker for the clinical assessment of osteoporosis](#)
- [3]: [Hydroxylysine - Wikipedia](#)
- [4]: [DL-5-Hydroxylysine 13204-98-3 - Sigma-Aldrich](#)
- [5]: [Lysine and novel hydroxylysine lipids in soil bacteria: amino acid ...](#)
- [6]: [Lysine - Wikipedia](#)
- [7]: [Urinary Hydroxyproline as an Inflammation-Independent Biomarker ...](#)
- [8]: [Hydroxylysine-Deficient Skin Collagen in a Patient with a... - JBJS](#)
- [9]: [A homozygous stop codon in the lysyl hydroxylase gene in two ...](#)
- [10]: [225400 - EHLERS-DANLOS SYNDROME, KYPHOSCOLIOTIC ...](#)
- [11]: [Ocular Ehlers-Danlos Syndrome With Normal Lysyl Hydroxylase ...](#)
- [12]: [Inherited human collagen lysyl hydroxylase deficiency: Ascorbic ...](#)
- [13]: [Seizures-intellectual disability due to hydroxylysinuria syndrome](#)
- [14]: [A Subtle Disease and a Dilemma: Can Cells Secrete Collagen That ...](#)
- [15]: [Hydroxylysine - Lab Results explained - HealthMatters.io](#)
- [16]: [Hydroxylysine - an overview | ScienceDirect Topics](#)
- [17]: [L-Hydroxyproline 2025: Benefits, Uses & Top Supplement](#)
- [18]: [L-Hydroxyproline 2025: Benefits, Uses & Top Supplement](#)
- [19]: [Proteomics Analysis of Lysine Hydroxylation and Cross-Linking of ...](#)
- [20]: [5-Hydroxylysine Analysis Service - Creative Proteomics](#)
- [21]: [Hydroxylysine - an overview | ScienceDirect Topics](#)
- [22]: [A Heritable Disorder of Connective Tissue: Hydroxylysine-Deficient ...](#)
- [23]: [4-Hydroxylysine | C6H14N2O3 | CID 550919 - PubChem](#)
- [24]: [Hydroxyproline - Wikipedia](#)
- [25]: [Disorder of lysine and hydroxylysine metabolism - Orphanet](#)
- [undefined]: [5-Hydroxylysine | C6H14N2O3 | CID 3032849 - PubChem](#)