International Journal of Culture and Modernity
ISSN 2697-2131, **Volume 36 | Jan-2024**
https://ijcm.academicjournal.io

Unveiling Nephrotic Syndrome: Causes, Pathogenesis, and Treatment

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ANNOTATION

This article explores Nephrotic Syndrome (NS), examining its diverse causes, pathophysiology, and tailored management strategies. With a focus on the Kidney Disease Improving Global Outcomes (KDIGO) guidance, the discussion navigates through complications, emphasizing the importance of individualized approaches for optimal outcomes.

KEYWORDS: Nephrotic Syndrome, causes, pathophysiology, KDIGO, management, complications.

Introduction: Nephrotic syndrome (NS) is a clinical syndrome characterized by massive proteinuria leading to hypoalbuminemia, resulting in hyperlipidemia, edema, and various complications. It is caused by increased permeability through the damaged basement membrane in the renal glomerulus, especially due to infections or thromboembolic events. NS can result from intrinsic renal diseases or be secondary to congenital infections, diabetes, systemic lupus erythematosus, neoplasia, or certain drug use. Nephrotic-range proteinuria is defined as the urinary loss of 3 grams or more of proteins per 24 hours or the presence of 2 g of protein per gram of urinary creatinine in a single spot urine sample. This proteinuria can also occur in systemic diseases like amyloidosis. The disorder can affect people of all ages. In most children, the first sign is facial swelling, while adults usually present with dependent edema. NS can occur in both genders and any race, either in a typical form or with nephritic syndrome, indicating glomerular inflammation leading to hematuria and impaired renal function.

Etiology: Common primary causes include intrinsic kidney diseases such as membranous nephropathy, minimal-change nephropathy, and focal glomerulosclerosis. Secondary causes may involve systemic diseases like lupus erythematosus, diabetes mellitus, and amyloidosis. Congenital/hereditary focal glomerulosclerosis may result from genetic mutations in podocyte proteins. Infectious diseases, allergic reactions, insect bites, vaccinations, and drug abuse, including heroin, can also trigger NS.

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Pathophysiology: Glomerular capillaries are lined by fenestrated endothelium, sitting on the glomerular basement membrane covered by podocytes. Destruction of podocytes leads to irreversible glomerular damage. The loss of plasma albumin through the glomerular filtration barrier is less than 0.1% in a healthy person. Proteinuria results from damage to the glomerular basement membrane, endothelial surface, or podocytes. Albuminuria, accounting for 85%, is associated with a generalized defect in glomerular permeability, causing nonselective proteinuria. Two hypotheses explain edema occurrence in NS. The under fill hypothesis links albuminuria to hypoalbuminemia, reducing plasma colloid osmotic pressure, leading to increased trans capillary filtration of water. The overfill hypothesis suggests intrinsic tubular defects leading to sodium retention and increased interstitial oncotic pressure. Different glomerulonephritides cause NS with varying histopathological features. Minimal change disease is common in childhood, while focal segmental glomerulosclerosis accounts for 10-15% of cases. Membranoproliferative glomerulonephritis involves immune complex deposition, and membranous glomerulonephritis is more common in adults. The first sign in children is facial swelling, progressing to generalized edema. Adults may present with dependent edema. Additional features vary based on the cause, and renal function impairment may lead to anemia, hypertension, or other complications.

Evaluation: Urine tests reveal nephrotic-range proteinuria, and urinalysis may show various casts. Blood tests indicate low serum albumin, and serologic studies help identify secondary causes. Ultrasonography demonstrates renal echogenicity, and renal biopsy is indicated in specific cases.

Treatment: This passage discusses the treatment and management of nephrotic syndrome. Before initiating corticosteroids, a comprehensive assessment is crucial, including monitoring height, weight, and blood pressure. Regular weight records aid in tracking edema changes, and physical examinations help detect infections and underlying systemic disorders. The specific treatment of nephrotic syndrome depends on its cause, leading to variations in management between adults and children. The Kidney Disease Improving Global Outcomes (KDIGO) issued guidance in 2012, providing recommendations for treating nephrotic syndrome.

In children, corticosteroids are primarily used for idiopathic nephrotic syndrome. Alternatives like cyclophosphamide, mycophenolate mofetil (MMF), calcineurin inhibitors, and levamisole may be necessary for those with frequent relapses or steroid-dependent nephrotic syndrome. Calcineurin inhibitors are the first-line choice for steroid-resistant nephrotic syndrome, with options like MMF or prolonged/intravenous pulse corticosteroids considered if there is no response. Rituximab, an anti-B cell antibody, serves as an effective steroid-sparing agent in pediatric cases. However, it may not achieve drug-free remission in children relying on both calcineurin inhibitors and steroids. Rituximab may also play a role in children with steroid-resistant disease. For children with complicated steroid-resistant nephrotic syndrome responding to rituximab, an additional treatment at B cell recovery may help maintain prolonged remission.

In adults, treatment varies based on the underlying cause. Minimal change nephropathy typically responds to prednisone. Lupus nephritis may require a combination of prednisone with cyclophosphamide or mycophenolate mofetil to induce remission. Secondary amyloidosis with nephrotic syndrome can improve with the anti-inflammatory management of the primary disease.

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Complications include metabolic consequences of proteinuria, infections, hypocalcemia, hyperlipidemia, hypercoagulability, hypovolemia, acute kidney injury, hypertension, edema, respiratory distress, and various infections.

In conclusion, nephrotic syndrome, marked by massive proteinuria, hypoalbuminemia, and edema, stems from glomerular damage with various etiologies, encompassing intrinsic kidney diseases to systemic conditions. Diagnosis involves urine and blood tests, with renal biopsy in specific cases. Treatment, tailored to the cause, includes corticosteroids, immunosuppressive agents, and targeted approaches. Prognosis varies; minimal change pathology often leads to remission, while focal-segmental glomerulosclerosis can progress to end-stage renal disease. Complications involve infections, metabolic issues, and cardiovascular risks, necessitating a comprehensive management approach for optimal outcomes.

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