

Clinical spectrum and pathological abnormalities of Paraprotein related Kidney disease - a single centre experience

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Introduction

- Spectrum of clinical manifestations in patients with paraproteinemia (MG) are diverse ranging from totally benign asymptomatic condition also called as monoclonal gammopathy of unknown significance (MGUS) to the malignant condition multiple myeloma (MM) characterized by either the impending or presence of end organ damage.
- Somewhere between this wide spectrum of clinical manifestations ranging from benign asymptomatic MGUS to malignant MM, an entity exists that affects kidneys where we can demonstrate paraprotein deposition in renal tissue but it does not meet the criteria to define MM.
- This entity is denoted by the term the Monoclonal gammopathy of renal significance (MGRS). Several pathogenic mechanisms can contribute to renal failure in patient with myeloma, some of which are the result of nephrotoxic Ig and some of which are independent of paraprotein deposition.
- We undertook this study to describe clinical manifestations, histopathological abnormalities in patients with paraprotein related kidney diseases

Methodology

- The study has been conducted in Department of Nephrology, Institute of Nephro-Urology, Victoria Hospital Campus Bangalore, India between January 2018 – July 2021
- A total of 60 patients were included in the study fulfilling following criteria:
 1. Biopsy proven myeloma cast nephropathy
 2. Renal biopsy findings suggestive of paraproteinemia such as immunofluorescence findings with light chain restriction as defined by International myeloma working group (IMWG)
- We collected clinical findings, laboratory findings and imaging abnormalities in prespecified proforma of these patients who were included in the study.

Results

- Of the 60 patients studied maximum number of patients were in 5th and 6th decade accounting for 27% (n=16) and 35% (n=21) (fig 1) respectively with mean age of 53.4 years.
- Male to female ratio in 3:1.
- Acute Kidney Injury (AKI) is the most common clinical presentation accounting for 35% of cases followed by Chronic Kidney Disease (CKD) which is 33%, Nephrotic syndrome in 11.6%, Rapidly Progressive Renal Failure (RPRF) presentation in 8.3%.

Renal histopathology found was Myeloma cast nephropathy 35%, followed by Amyloidosis AL which is the second most common renal lesion seen in 18.3% of cases (fig 3). 15% had Proliferative Glomerulonephritis with Monoclonal Immune-Deposition disease (PGNMID), 8.3% having non amyloid deposition disease. Monoclonal immunoglobulin Deposition Disease (MIDD) was seen in 10% patients.

47.5% of patients required dialysis at presentation

Fig 1

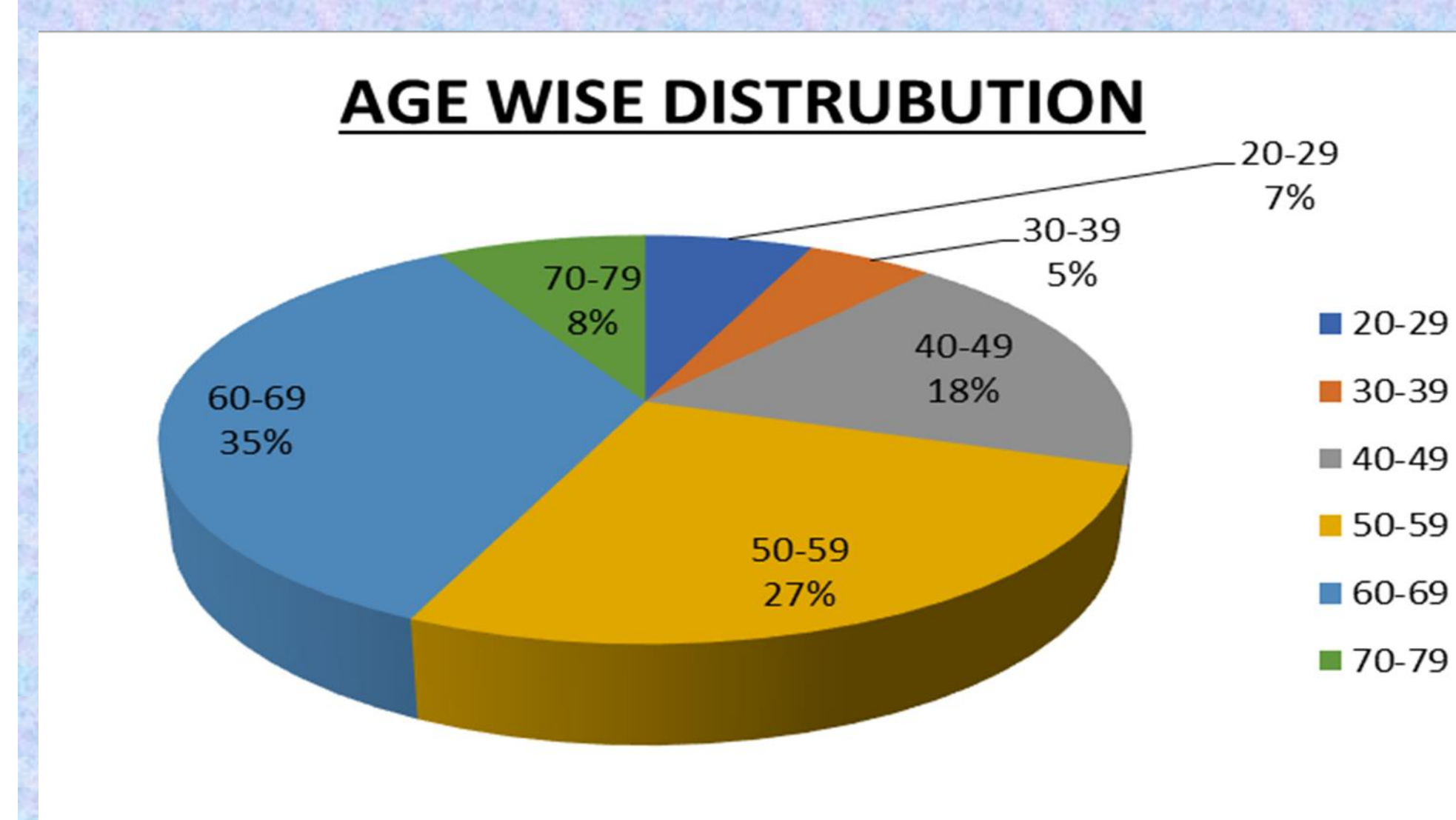


Fig 2

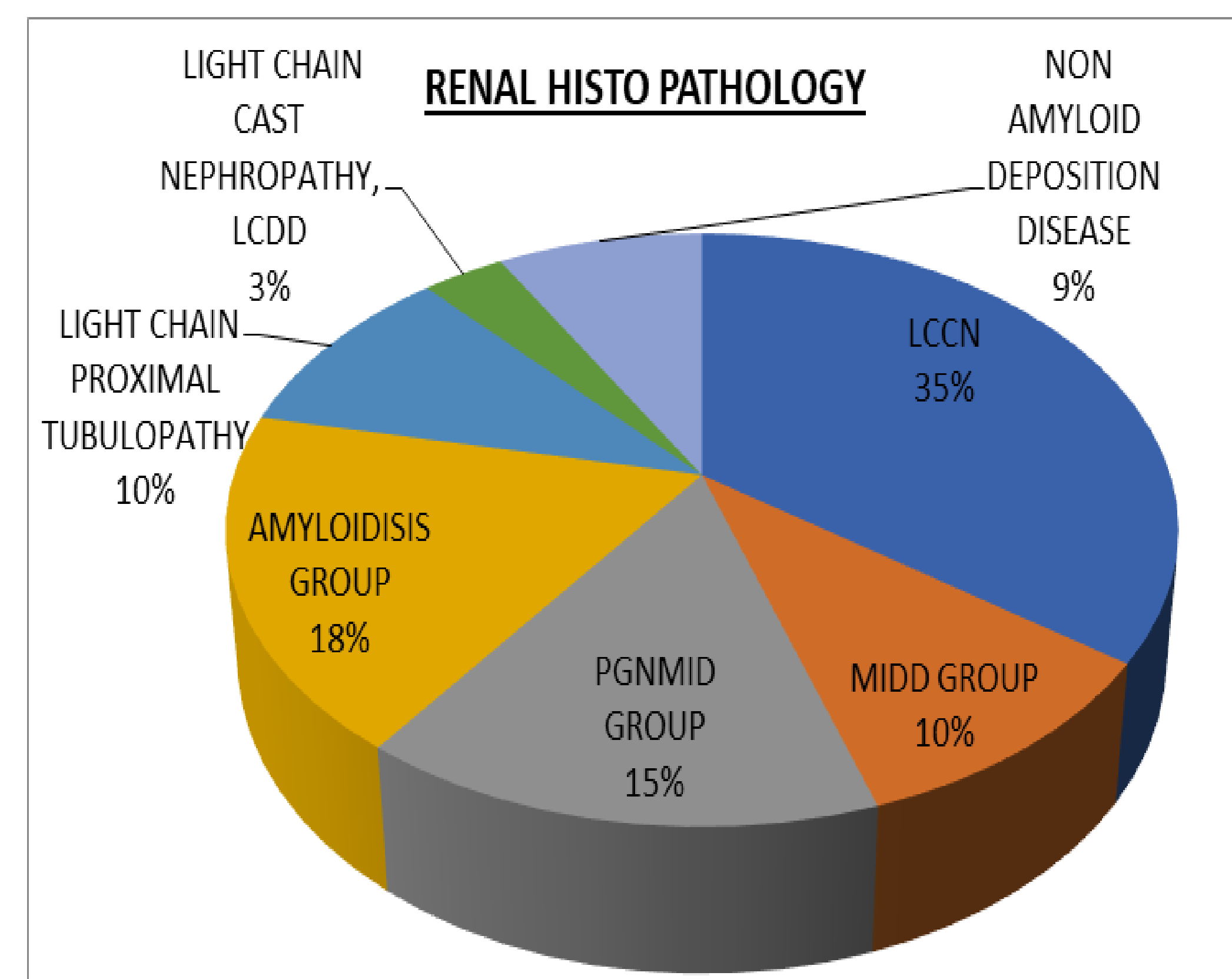
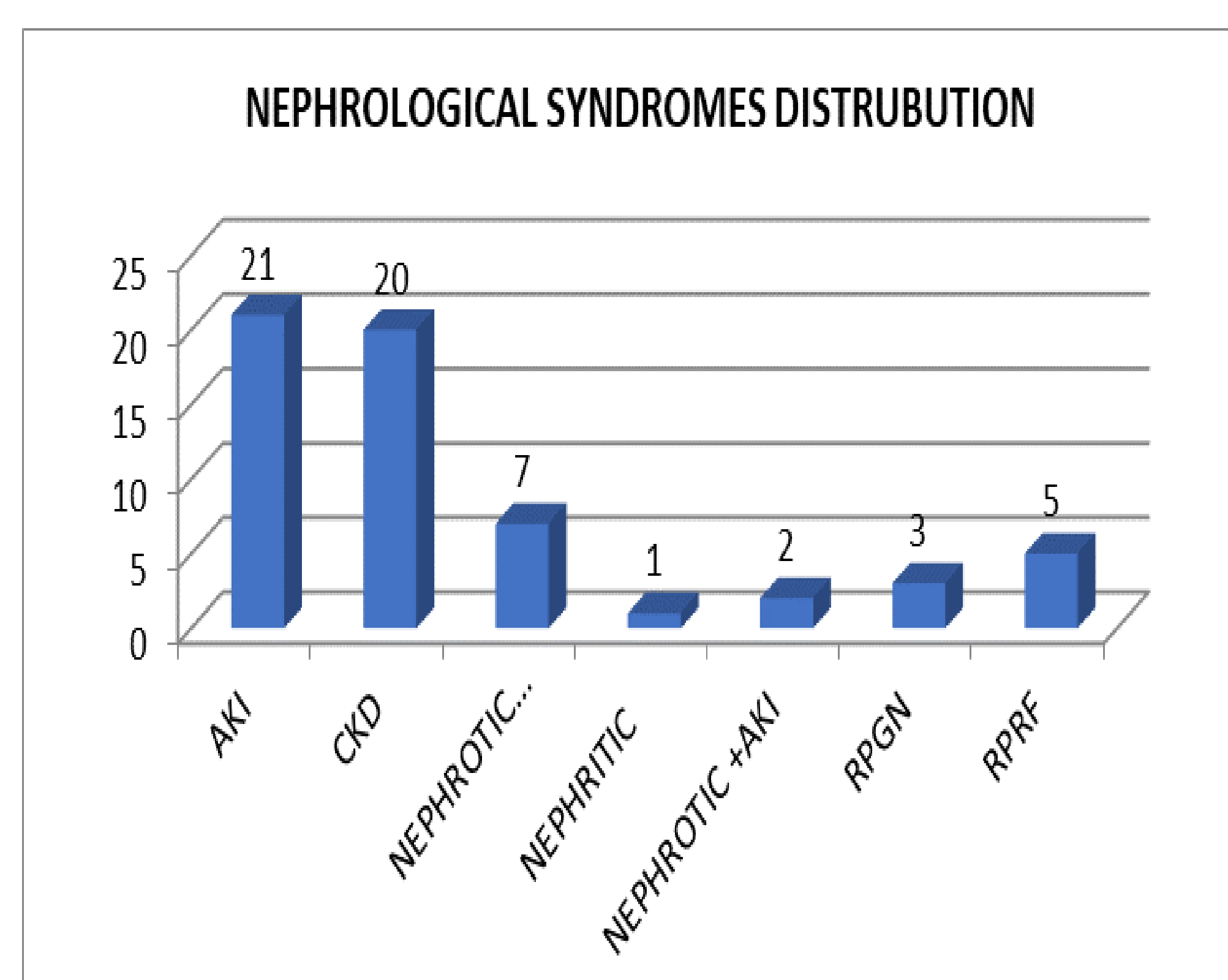


Fig 3



DISCUSSION

- PW sanders et al Thirty-six patients (2.6% of 1361 kidney specimens examined over 6 years) had evidence of monotypical light chain with or without concomitant heavy chain deposition. AL-amyloid was the most common renal lesion (39%), nonamyloid deposition occurred second most commonly (33%), and cast nephropathy ("myeloma kidney") was third most frequent (14%).
- Start DA, et al conducted a systematic study from 53 patients with plasma cell dyscrasia, 24 of whom had Bence Jones cast nephropathy (with large casts, often associated with giant cells and polymorphonuclear leukocytes), interstitial nephritis or fibrosis without cast nephropathy (20-30%), amyloidosis (10%), LCDD (5%), ATN (10%), Other 5%

Conclusion

- Paraproteinemia related kidney diseases are associated with diverse clinical manifestations. AKI was most common clinical presentation followed by CKD in our study.
- Among histopathological spectrum Myeloma cast nephropathy was most common pathological abnormality observed in our study followed by AL Amyloidosis.

References

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