

Renal cysts in Thin Basement Membrane Disease with proteinuria/renal function impairment

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Introduction

- Thin basement membrane disease (TBMD) is the main cause of long term microscopic hematuria.
- Around 60% of patients develop proteinuria, that reaches >0.5 g/d in approximately 10% of the patients.
- In some studies the presence of proteinuria increases the risk of hypertension and renal function impairment in TBMD.
- The pathogenic mechanisms responsible of the appearance of CKD in TBMD are presently unknown.

Aim of the Study:

- To describe the clinical, radiological, histological and laboratory findings at the onset and at the end of follow up in a cohort of patients diagnosed with TBMD who developed proteinuria over 0,5 g/24 h

Methods

- Descriptive, observational, retrospective study to analyze all our patients with TBMD and proteinuria over 0,5g/24h.
- Sixteen patients belonging to 12 families were included in the study. They were diagnosed between 1979 and 2007 and followed from January 1979 up to September 2012.
- Patients were diagnosed with TBMD according to the following criteria: Diagnostic renal biopsy findings; long term microscopic hematuria lacking identifiable origin and having three or more first line relatives with microscopic hematuria or first line relatives with biopsy-proven TBMD.
- Patient's medical records were reviewed for demographical information, clinical, radiological, laboratory and histological findings.

Results

Clinical characteristic at the start and end of follow up

N:16
10♂:6♀
Follow up: 198 (30-290) moths
12 families: 4 families with two patients, the rest just one

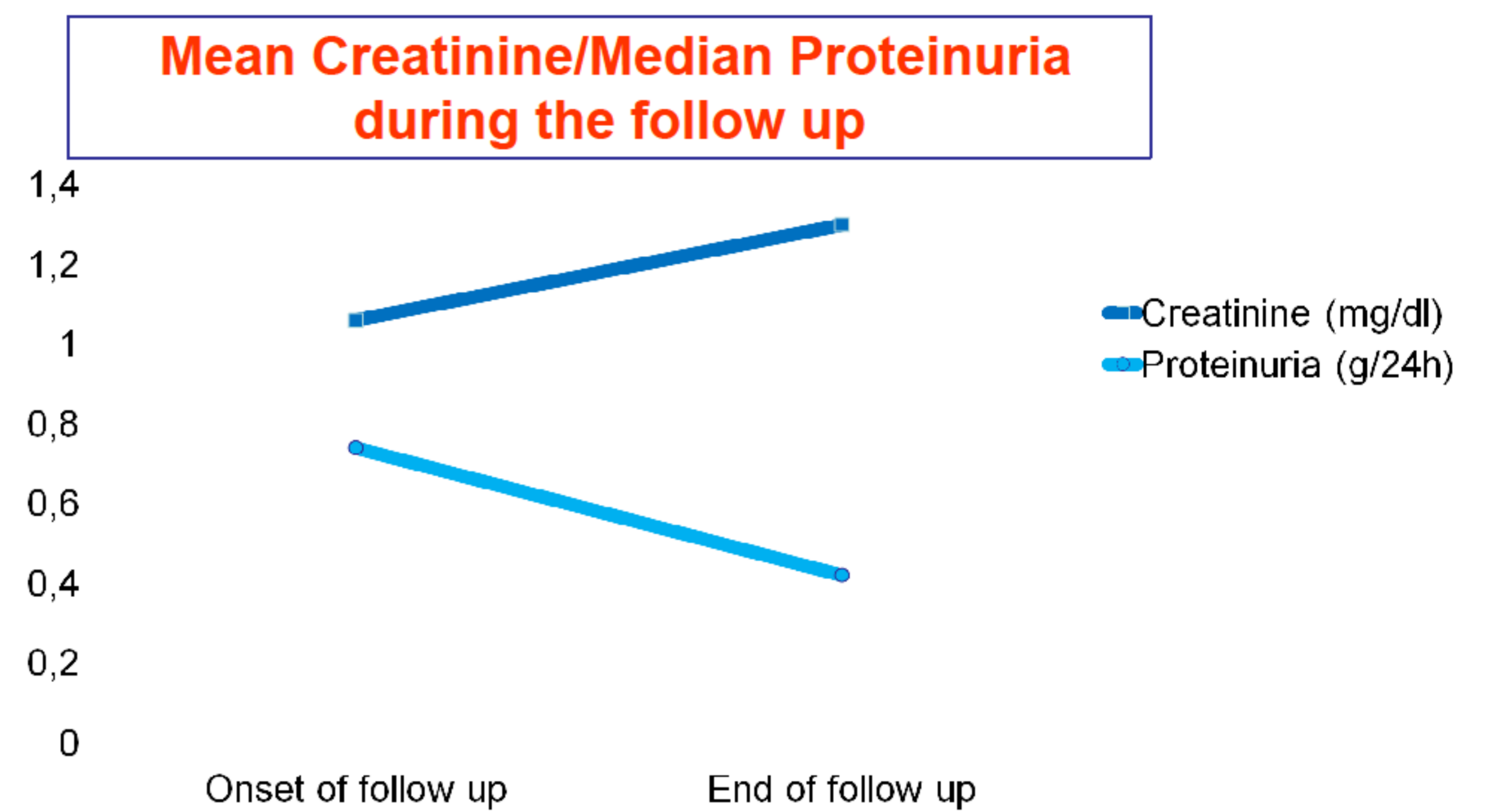
	Onset of follow up	End of follow up
Age	35,12±17,31 (7-62)	47,28 ±14,85
HTN	2 (12,5%)	11 (68,75%)
DM 2	0	1 (6,25%)
DL	1 (6,25%)	11 (68,75%)
Obese	4 (25%)	6 (37,5%)
Smoker	4 (25%)	4 (25%)
Lithiasis	4 (25%)	4 (25%)
Urinary infections	2 (12,5%)	2 (12,5%)
GFR below 60	3 (18,75%)	7 (43,75%)

HTN: Blood Hypertension ; DM 2: Diabetes mellitus 2; DL: Dislipemia; GFR: Glomerular filtrate rate (calculated by MRDR-4 y expressed in ml/min/1,73m²); Age: years

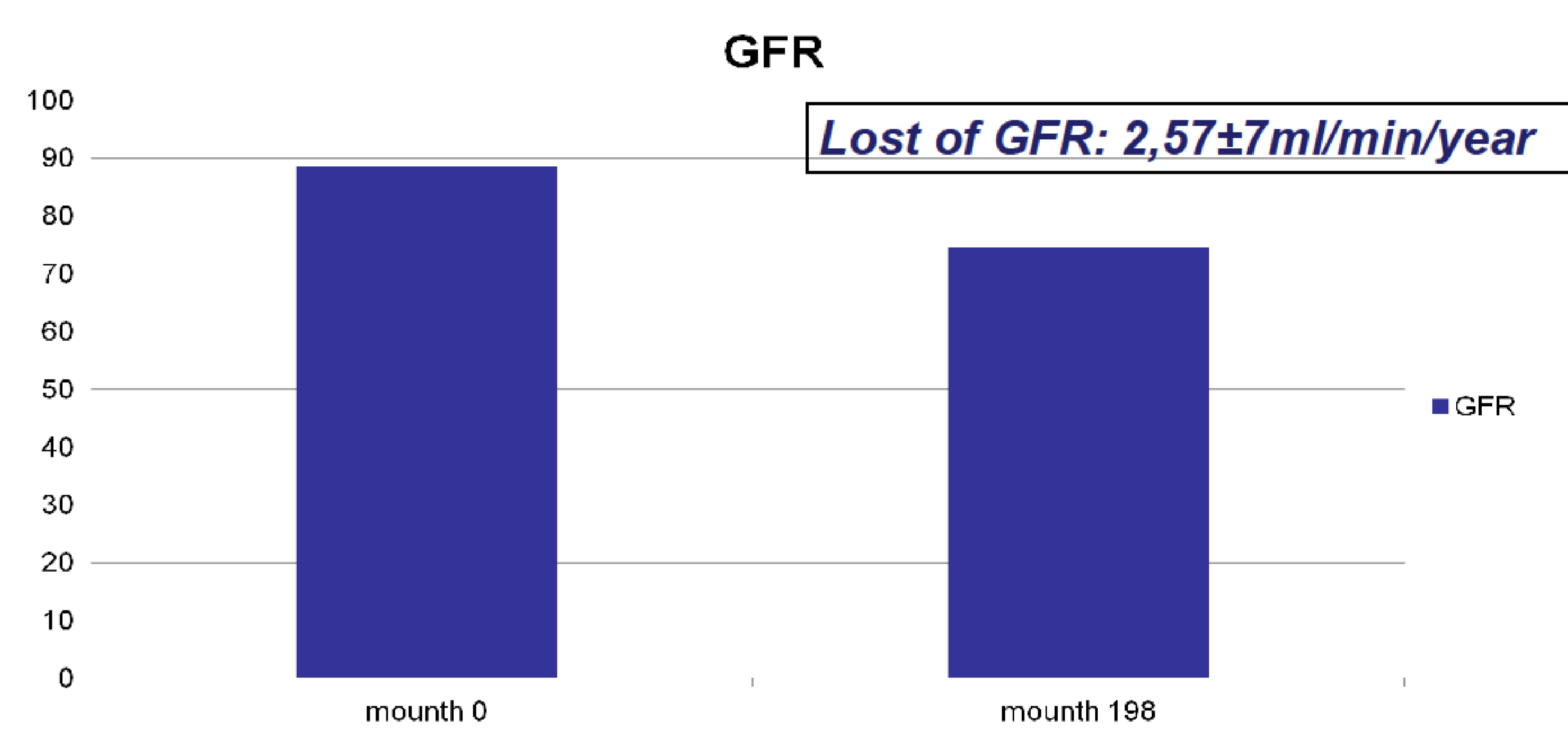
Conclusions

- Patients with TBMD and proteinuria >0.5 g/24hr can present a slowly progressive renal function decline. The proteinuria was well controlled after the start of the treatment with RAAS blockers. A significant proportion (>50%) exhibit bilateral renal cysts, whose possible genetic basis and influence on renal outcomes require further investigations.

Results (cont.)

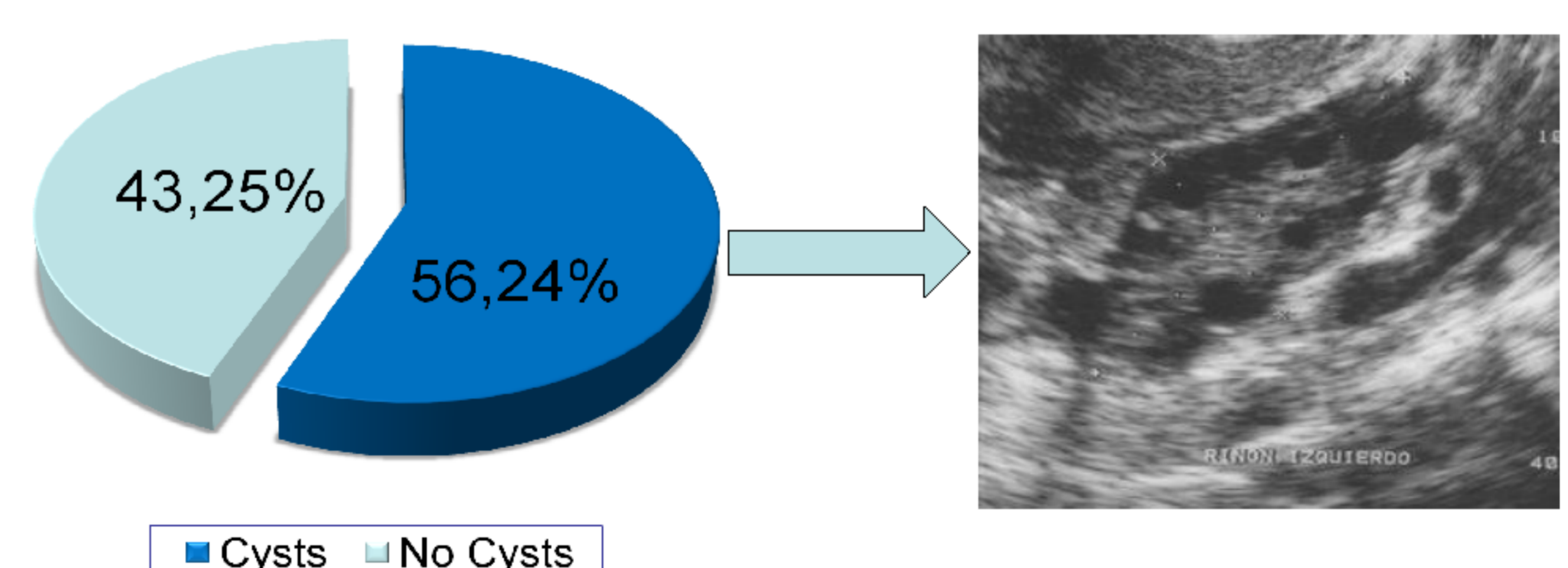


All the patients were treated with RAAS blockers



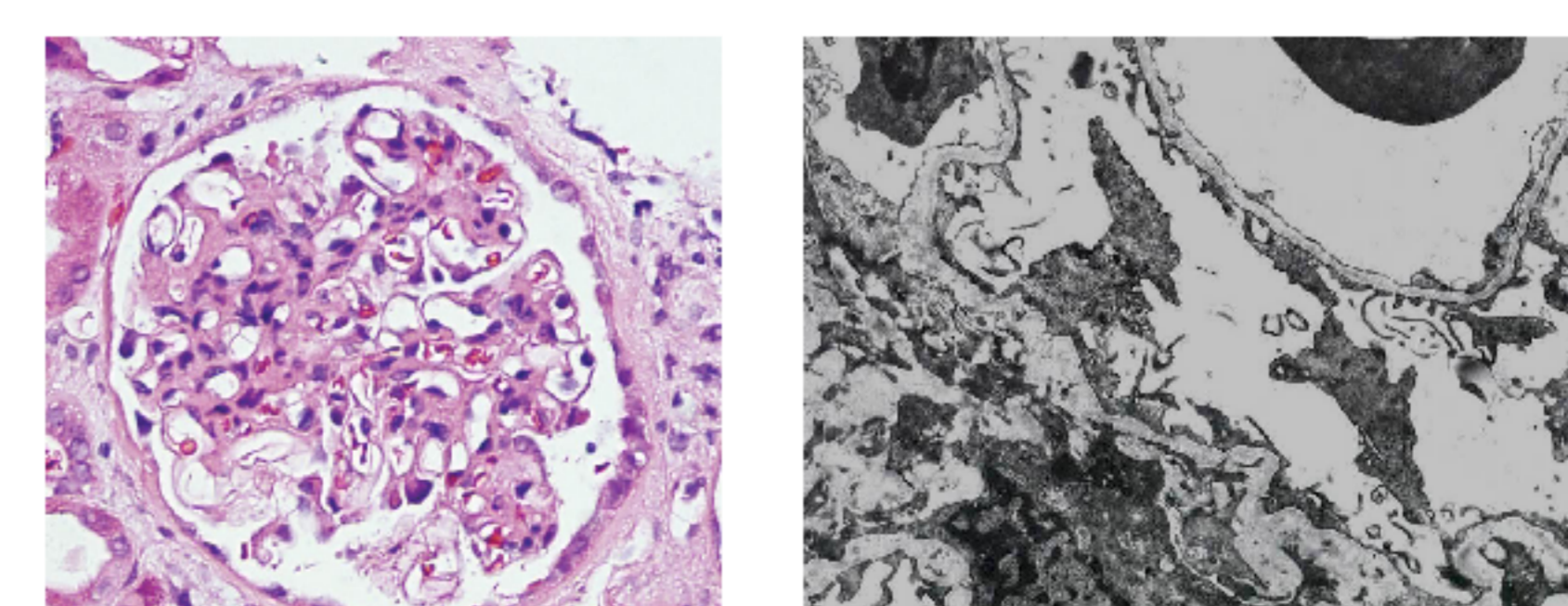
Glomerular Filtrate Rate (ml/min/1.73m ²)	Patients at the onset of follow up	Patients in the end of follow up
>90	7 (43,75%)	6 (37,5%)
60-90	6 (37,5%)	3 (18,75%)
45-59	1 (6,25%)	2 (12,5%)
<45	2(12,5%)	5 (31,25%)

Ultrasound findings



Normal sized kidneys
Bilateral cysts
Cysts size between 10-70 mm
Similar outcomes in the follow up

Histological finding



LM: Occasional mild mesangial cellular proliferation and matrix. Low glomerular sclerosis
ME: Uniform thinning of the glomerular basement membrane under 250 nm. NO lamination

