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LETTER TO THE EDITORS

Response to: Urine bikunin and kidney involvement in Fabry disease

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Dear editors,
We thank Drs. Lepedda and Sechi for their critical analysis of our article [1].

We agree with their comments about the need for identifying renal biomarkers to detect kidney impairment in pediatric Fabry disease (FD) patients, with the advantages of precocious diagnosis and therapies. The obsolete biomarker, serum creatinine, does not detect kidney damage precociously. Moreover, glomerular filtration rate, proteinuria, and albuminuria are unable to discover sub-clinical kidney dysfunction. These biomarkers could provide false-negative results, underestimating the incidence and prevalence of kidney damage in FD patients, especially when the functional renal reserve is not entirely lost. However, we believe that the suggestion of bikunin as a kidney biomarker does not help achieve this target.

The authors cited their data published about 10 years ago, evaluating urine bikunin in 24 adult patients, with a suggested diagnostic role in FD patients [2]. Urine bikunin did not correlate with serum creatinine, and pathological proteinuria and overt kidney damage characterized the enrolled patients. What was the role of this chondroitin sulfate proteoglycan in FD patients? Can the data obtained in adults be transferred to the pediatric field?

Since then, no other evidence has strengthened these results or explained the origin of urine bikunin and the mechanisms inducing elevated urine levels in FD patients. The same authors underlined these doubts in their work. Moreover, which additional diagnostic or prognostic information could this biomarker add?

A recent work assessed bikunin precursor levels in treated adult FD patients, revealing increased values in patients with kidney disease progression. However, this biomarker did not show the sensitivity of other markers, such as nephrin or podocalyxin, cited in our review, indicating that this protein was altered in advanced kidney disease only [3].

Lepedda and Sechi criticized the exclusion of urinary bikunin from our review section entitled "Kidney damage assessment in FD: from biomarkers to kidney biopsy" as a potential marker with clinical utility. Their study was the first and only report revealing high urine bikunin levels in adult FD patients.

In our opinion, literature data have not confirmed, in the last decade, bikunin as a promising biomarker in FD patients, justifying the lack of citation in our educational work, which should attract the interest of nephrologists and pediatricians regarding the precocious diagnosis of renal involvement in FD patients.

Declarations

Conflict of interest The authors declare no competing interests.

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