



# The Baseline Serum IgG4/IgG3 Ratio Might be a Suitable Marker for Predicting Favorable Treatment Response in Patients With Idiopathic Retroperitoneal Fibrosis-associated Hydronephrosis

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Retroperitoneal fibrosis (RPF) is a rare disease featuring the proliferation of fibrous tissue in the retroperitoneum, the compartment of the body containing the kidneys, aorta, renal tract, and various other structures. It may present with lower back pain, kidney failure, hypertension, deep vein thrombosis, and other obstructive symptoms [1]. Ureteral involvement has been reported in 80%~100% of the cases at presentation, occasionally causing hydronephrosis [2]. Hydronephrosis is a common complication of RPF and it may lead to poor outcome without appropriate intervention. RPF is categorized as secondary or idiopathic causes [3]. Secondary causes of RPF include infection, drug use, malignancy, radiation therapy and abdominal operation. Idiopathic RPF (iRPF) accounts for two-thirds of the RPF cases and can be divided into two groups: IgG4-related RPF and non-IgG4-related RPF [4]. Histologic confirmation would be great to diagnose IgG4-related RPF because treatment strategy for IgG4 related disease is well-defined. However, when we manage the patients with RPF in real clinical practice, it is very difficult to obtain sufficient tissues owing to the location of fibrosis and high risks associated with open biopsy. In addition, there have been no validated clinical parameters for assessing outcome of iRPF [5-7].

A recent paper by Choi et al. [8] published in *Journal of Rheumatic Diseases* investigated for the first time the predictive factors for hydronephrosis-associated outcomes among iRPF patients using clinical profiles and serum

IgG subclasses. They retrospectively included 18 iRPF patients with hydronephrosis. Hydronephrosis improvement was assessed by image taken 6 months after diagnosis. On follow-up images, 8 patients (44.4%) showed improvement in hydronephrosis. Patients with improvement more frequently had reverse serum IgG4/IgG ratio (87.5% vs. 30%,  $p=0.025$ ), abdominal aorta involvement (87.5% vs. 30%,  $p=0.025$ ) and glucocorticoid treatment (87.5% vs. 30%,  $p=0.025$ ) than those without improvement. The proportion of elevated serum IgG4 level did not differ between the two groups. Interestingly, even in the 14 cases with normal serum IgG4 levels, it was found that reverse serum IgG4/IgG3 ratio was more frequent in patients with improvement than in those without improvement (83.3% vs. 12.5%,  $p=0.026$ ). So, Choi et al. [8] could draw a conclusion that baseline serum IgG4/IgG3 ratio might be a suitable serologic marker for predicting good treatment response in iRPF patients with hydronephrosis.

The study by Choi et al. [8] has two major limitations. One is the limited number of cases that was far from the strong statistical power. The other is the fact that tissue biopsy was not performed in all the cases. Nevertheless, clinical predictor for favorable hydronephrosis-associated outcomes in itself might be invaluable in that there been no validated clinical parameters for assessing outcome of iRPF. Thus, the rheumatologists could consider the application of baseline serum IgG4/IgG3 ratio as predictor for

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favorable response of hydronephrosis when managing the iRPF patients.

### CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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