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# A Biomarker Cyst Fraction in Autosomal Dominant Polycystic Kidney Disease

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## Introduction

The most common monogenic kidney disease is autosomal dominant polycystic kidney disease. Patients at high risk of severe disease progression should be identified early so that supportive and therapeutic measures can be implemented. The glomerular filtration rate, on the other hand, may remain within normal limits for decades before decline begins, making it a late indicator of rapid progression. Kidney volumetry is frequently used in clinical practise to determine disease severity. Due to poor prognostic accuracy, additional imaging markers are being sought to improve outcome prediction in ADPKD, but data from clinical cohorts is still scarce. In this study, we looked at cyst fraction as one of these parameters in 142 ADPKD patients.

The most common genetic cause of kidney failure is autosomal dominant polycystic kidney disease, which is characterised by the progressive enlargement of multiple bilateral renal cysts and a decline in kidney function. In ADPKD, the loss of kidney parenchyma due to cyst growth is continuous and irreversible. The glomerular filtration rate, on the other hand, often remains within a normal range for many years until a more rapid decline in kidney function occurs from the fourth decade of life onward. There are currently no treatment options for ADPKD. Current treatments primarily aim to prevent renal cytogenesis and the resulting destruction of the kidney parenchyma.

## Description

Importantly, the beneficial effect is time dependent, making early identification of patients at high risk of severe disease progression a valuable asset. Precise outcome prediction allows for better patient counselling and prevents patients with slow disease progression from being exposed to potential side effects. Various prognostic factors for ADPKD have entered clinical practise to date, including genetic, clinical, and imaging data. Total kidney volume, as measured by MRI-based volumetry, is the most commonly used biomarker in ADPKD and, as such, can be regarded as the gold standard for predicting outcome [1].

However, simply using volume ignores the fact that a single large cyst can dominate this parameter with little impact on kidney integrity. As a result, knowing how much normal parenchyma has been replaced by cysts appears to be an important addition to the volume itself. Cyst fraction is not part of the clinical routine, owing to a lack of data on its potential and the time-consuming nature of its evaluation. As a result, more evidence is urgently needed to uncover the potential of this parameter. The current study compared cyst fraction to TKV, correlating both parameters to kidney function in a group of 142 ADPKD patients.

Kidney volume was calculated using semi-automatic volumetric segmentation and manual editing in the IntelliSpace Discovery software. The majority of the MRI images were obtained by combining a T2-weighted Spectral Presaturation with an Inversion Recovery sequence. Other sequences, such as Fluid Attenuated Inversion Recovery, were used if none were available. Following the completion of the TKV measurement, the cyst volume was calculated using ISD's "Threshold Segmentation" plugin. The AV-Score of the brightest area of the kidney parenchyma was measured for this, and this served as the lower masking threshold.

The gold standard for patient selection of therapeutic measures in ADPKD has become prognostic imaging parameters. An increase in kidney volume is associated with disease severity. TKV height adjustments revealed that even a single examination of the htTKV can predict the risk of disease progression. However, because kidney atrophy/fibrosis is a possibility, kidney volume alone is not always informative. Furthermore, single dominant cysts have a high impact on TKV while having little effect on the parenchyma [2-5].

## Conclusion

This study adds to the existing evidence on the potential of cyst-fraction, providing valuable information to TKV. These findings should now serve as the foundation for future studies focusing on predicting future eGFR in larger cohorts and in combination with other imaging and non-imaging biomarkers. Furthermore, it will be critical to continue developing tools that allow for simple measurement of the cyst fraction in order to enable clinical use in the future.

## Acknowledgement

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## Conflict of Interest

There are no conflicts of interest by author.

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