

# Sequence of Surgical Approach in a Patient with Autosomal Dominant Polycystic Kidney Disease Treated with Renal Transplantation

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## ABSTRACT

**Introduction:** Describe the surgical sequence in a 55-year-old woman with an Autosomal Dominant Polycystic Kidney (ADPKD)

**Case Presentation:** We performed three surgical procedures in a woman with Chronic Kidney Disease (CKD) secondary to ADPKD of 13 years of evolution from June to November 2023 which are as follows: Resection of 2 liver cysts and Laparoscopic cholecystectomy, Related living donor renal transplant, and Bilateral open nephrectomy, all of them with the use of innovative technology.

**Result:** The first procedure was performed for risk reduction and two cysts of 3.5 cm x 1.6 cm x 1.5 cm and 3.3 cm x 2.0 cm x 1.4 cm and chronic cholecystitis were reported. The second one was performed one month later; the patient had previous creatinine values of 4.73. The graft was positioned in the right iliac fossa where end-to-side anastomosis of renal vessels was performed with external iliac vessels and uretero-vesical anastomosis was executed with the Lich-Gregoir technique; the total ischemia time was 109 minutes and the kidney presented immediate function. At 48 hours, a Doppler ultrasound was performed, which reported a renal artery resistance index of 0.5, with homogeneous cortical perfusion, without collections, vessels, and ureter without stenosis or thrombosis; therefore, the patient was discharged 5 days after the transplant with optimal graft function and creatinine levels of 1.75. In the last procedure, two kidneys of 1.6 kg each were obtained and pathology reported adult polycystic disease with interstitial fibrosis, chronic inflammation, dystrophic calcification, tubular and glomerular atrophy. At the moment she has stable graft function and went from having creatinine levels of 4.73 to 1.2 and her glomerular filtration rate went from 9.68 to 50.84.

**Conclusion:** In our experience, performing liver cyst resection at the beginning

reduced the possibility of risks in the transplant, which was anticipated for better graft acceptance; in addition, high specialty surgery with the use of ICG and special sutures reduced ischemia times and provided greater safety to the procedure. Subsequently, a nephrectomy was performed and after this procedure there were no complications and graft function did not diminish. So, our initial experience can serve as a guide for future studies in search of generating a sequential surgical treatment of patients with PKD for adequate graft acceptance and improvement in quality of life.

**Keywords:** Renal Transplantation • Living Donor • Nephrectomy • Indocyanine Green • Insufflation Systems

## INTRODUCTION

Autosomal Dominant Polycystic Kidney Disease (ADPKD), first described in 1841, is the fourth leading cause of Chronic Kidney Disease (CKD) [1]. Its origin is a product of mutation of 2 chromosomes, the PKD1 gene on chromosome 16 which causes 85% of cases, and PKD2 on chromosome 4 which causes the other 15% of cases [1]. In the United States, it is considered to have an incidence of approximately 5000 cases per year [1].

ADPKD is a disease that can cause renal manifestations such as nocturia, pollakiuria, arterial hypertension, or extrarenal data such as polycystic liver disease or valvular prolapse [1].

Diagnosis can be made with imaging studies such as Ultrasound (USG) of the abdomen or abdominal Computed Tomography (CT), and screening studies are also recommended in individuals with family members with the diagnosis [1]. Renal transplantation is the best replacement therapy following loss of renal function [1].

The number of patients with a diagnosis of ADPKD will require native nephrectomy at some point in their lives, either due to recurrent infections, renal pain, urolithiasis, suspected malignancy, or decreased vital space for placement or permanence of the renal graft [2-4], and there is debate about the moment at which to perform it [5]. Several studies do not recommend pre-transplant nephrectomy because it is associated with greater morbidity and mortality compared to post-transplant patients [2-6].

Nephrectomy at the time of transplantation is also not used in a standardized way because of the risk of generating potential damage to the graft in addition to decreasing the protective factor of residual renal function and performing another unnecessary surgical procedure that can lead to graft rejection [5]. Likewise, it is mentioned that performing native nephrectomy does not affect the functioning of the renal graft nor does it increase the immunological risk [7-8].

Regarding the prognosis of patients with ADPKD after renal transplantation and nephrectomy, they have a better prognosis than patients who do not have ADPKD and received renal transplantation for other causes such as

diabetes mellitus, hypertension or glomerulonephritis, because in the former there is less recurrence of native disease which may increase the risk of graft loss, also, older donors or cadaveric donors increase the risk of graft failure [5].

We will present the case of a 55-year-old woman diagnosed with CKD KDIGO (Kidney Disease: Improving Global Outcomes) 5 secondary to ADPKD who underwent sequential surgical treatment starting with cholecystectomy and liver cyst resection, followed by renal transplantation and ending with bilateral native nephrectomy.

## CASE PRESENTATION

A 55-year-old woman with a diagnosis of CKD KDIGO 5 secondary to ADPKD, hemotype B+, physical activity swimming, and previous surgical total abdominal hysterectomy. Weight: 73 kg, Height: 1.63 m. BMI: 27.54.

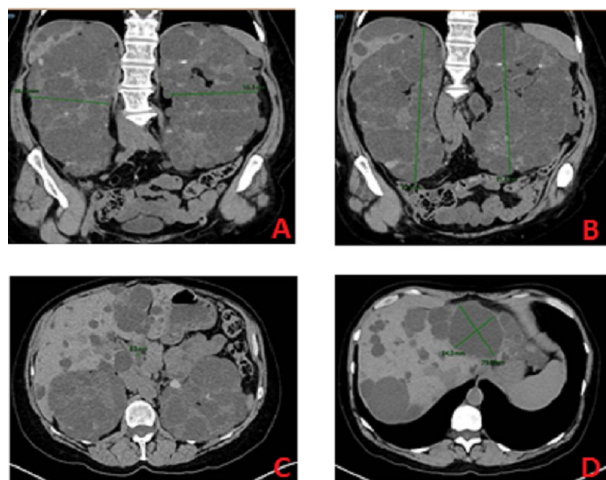
The patient's clinical picture consisted of lumbar pain 8/10 and recurrent urinary tract infection (UTI) with medical treatment. The diagnosis of ADPKD was made at the age of 37 years after presenting recurrent UTI and CKD was diagnosed 2 years later when the need for renal replacement therapy was indicated.

In January 2023, the organ transplantation protocol with medical equipment began. An abdominal Computed Tomography (CT) scan was performed which reported liver and kidneys with lobulated contours, with increased dimensions, multiple rounded images, and well-defined regular contours of liquid density (Figure 1).

A thyroid Ultrasound (USG) was performed, reporting a symmetrical and homogeneous thyroid gland with heterogeneous parenchyma due to the presence of multiple nodular images, and a chest CT scan was performed, reporting a right pulmonary nodule with an apical granulomatous appearance.

Laboratory studies reported a glomerular filtration rate of 9.9 ml/min/1.73 with alteration of all urinary function parameters (Table 1).

The complete protocol was presented to the transplant In



**Figure 1:** A: Abdominal CT scan with transverse measurement of polycystic kidneys 9 cm right and 10 cm left; B: CT of the abdomen with longitudinal measurement of polycystic kidneys 22.9 cm right and 21.6 cm left; C: CT scan with cystic images in the liver; D: CT scan with cystic images in the liver.

**Table 1:** Urinary Function Tests.

Parameter	Result	Units	Reference values
Creatinine	4.66	(mg/dL)	0.70-1.30
BUN	69.1	(mg/dL)	8.4-25.7
BUN/Creatinine	14	(mg/dL)	7.0-24.0
Urea	147	(mg/dL)	10.7-53.5
Glomerular Filtration Rate	9.9	(mL/min/1.73 m <sup>2</sup> )	70.00-110.00

BUN: Blood Urea Nitrogen; mg/dl: milligrams per deciliter; mL/min/1.73 m<sup>2</sup>: milliliters per minute per 1.73 square meters

June 2023, the first surgical procedure was performed for risk reduction, consisting of a laparoscopic cholecystectomy and resection of two hepatic cysts. Pathology reported the presence of two giant liver cysts of 3.5 cm x 1.6 cm x 1.8 cm and 3.3 cm x 2 cm x 1.14 cm and a gallbladder of 7.0 cm x 2.6 cm x 0.4 cm with edema, vascular congestion, and scarce mononuclear inflammatory infiltrate.

Subsequently, the transplantation protocol and compatibility tests with the related living donor (son) were completed and blood typing of ABO and HLA groups (HLA-A, -B, and -DR) was performed.

The complete protocol was presented to the Hospital Angeles Clínica Londres transplant committee and was accepted as a recipient of the left kidney from a related person. Two months after the first procedure, renal transplantation was performed simultaneously with the laparoscopic living donor nephrectomy. The transplant

consisted of open surgery using Indocyanine Green (ICG) and expanded Polytetrafluoroethylene (PTFE) sutures.

After general anesthesia, orotracheal intubation was performed, a foley tube was placed and surgical marking was performed for graft placement in the right iliac fossa (Figure 2).



**Figure 2:** Surgical marking.

ICG was injected intravenously on three occasions: 2 milliliters (ml) at the time of anesthetic induction, at 20 minutes, and 40 minutes respectively, and the patient was placed in supine decubitus. Support areas were protected and a compressive bandage was applied to the lower extremities.

The graft was placed in the right iliac fossa where end-to-side anastomosis of the renal vein with the right external iliac vein was performed with CV-6 caliber suture (Table 2) and of the single renal artery with the right external iliac artery with Continuous Suture Standard Technique with CV-7 caliber suture (Table 2), subsequently, Intraoperative perfusion was verified with the use of the ICG (Figure 3).



**Figure 3:** Verification of graft perfusion with ICG.

The uretero-vesical anastomosis was performed with the Lich-Gregoir technique and presented immediate function. Total ischemia time was 109 minutes (Table 3). There was no blood leakage at vessel stripping and the bleeding quantification was 150 ml. A Doppler Ultrasound of the renal graft was performed at 48 hours and reported a renal artery resistance index of 0.5 with laminar flow, visualization of vessels and ureter without stenosis or thrombosis, homogeneous cortical perfusion, no fluid or blood collections, and normal echogenicity.

**Table 2:** Specifications of PTFE sutures used during the procedure.

Caliber	Length	Needle	Needle:-Thread	Measure (USP)
CV-6	61 cm (24 in)	Conical 3/8 9 mm	1:01	5-0
CV-7	61 cm (24 in)	Conical 3/8 13 mm	1.6:1	6-0
CV-8	76 cm (30 in)	Conical 3/8 9 mm	1.9:1	7-0
USP: United States Pharmacopeia				

**Table 3:** Ischemia times.

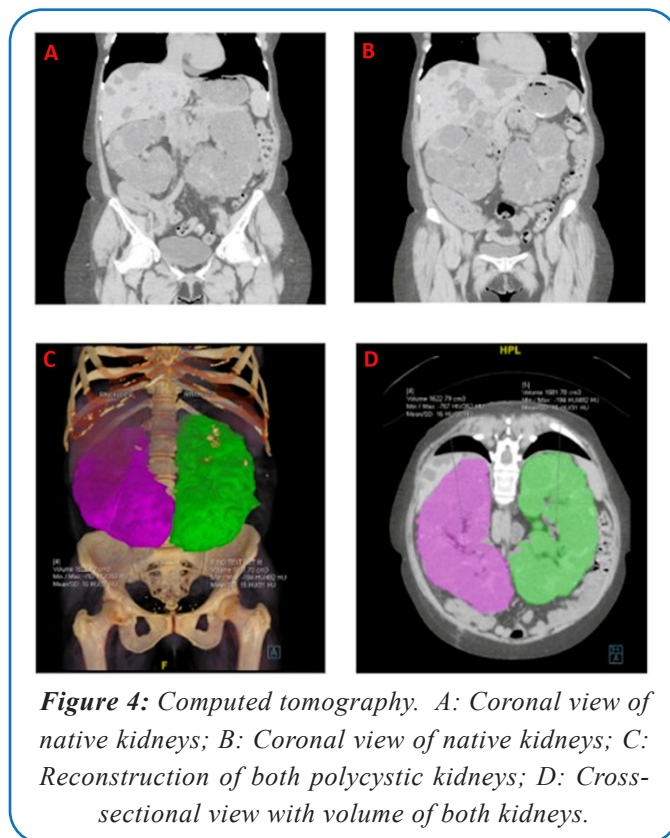
Ischemia time	Start	End	Total:
Hot ischemia	16:26:20 hours	16:29:28 hours	03 minutes 08 seconds
Cold ischemia	16:29:28 hours	17:21 hours.	51 minutes 32 seconds
Warm ischemia	17:21 hours	18:15:51 hours.	54 minutes 51 seconds
<b>Total</b>			01 hour 49 minutes 31 seconds

Immunosuppressive treatment was started and the follow-up and evaluation protocol was continued. Due to the persistence of lumbar pain, a CT scan was performed, which showed multiple ovoid images of regular borders in the liver, the largest of 6 cm x 7 cm, and both kidneys were enlarged, the right one with a longitudinal diameter of 25 cm and the left one of 23 cm with heterogeneous parenchyma and the presence of multiple cystic images. Similarly, evidence of compression by the ipsilateral native kidney was identified in the renal graft (Figure 4).

Therefore, it was decided to perform an open bilateral nephrectomy to reduce the risk of infection of the native kidneys and to prevent lesions due to loss of the vital space of the renal graft.

After general anesthesia, orotracheal intubation was performed, a foley tube was placed and the midline surgical marking for laparotomy was performed (Figure 5).

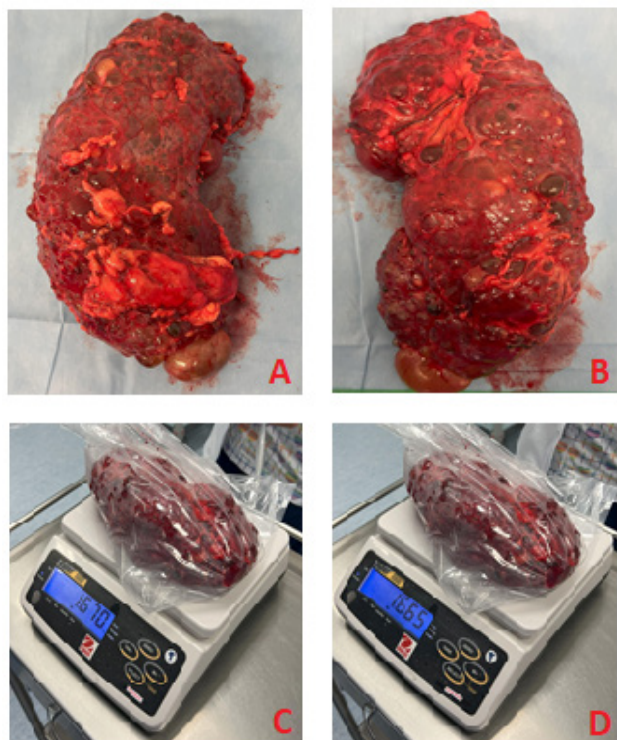
ICG was injected intravenously in a 2 ml injection at the time of anesthetic induction. Support areas were



protected, and a compressive bandage was applied to the lower extremities. The patient was then placed in the supine position.

The right kidney weighing 1.670 kg (kilograms) and measuring 30 cm in length was extracted first, followed by the left kidney which weighed 1.665 kg and measured 29 cm in length (Figure 6).





**Figure 6:** A: Right kidney; B: Left kidney; C: Weight of 1,670 kg of right kidney; D: Weight of 1,665 kg of left kidney.

The surgical procedure was completed without any complication and after observation and laboratory control, the patient was discharged after 72 hours without incident. The pathology report refers to the presence of adult polycystic disease with interstitial fibrosis, chronic inflammation, dystrophic calcification, tubular and glomerular atrophy, and surgical margins without histological alterations.

## DISCUSSION

ADPKD is a relatively low-incidence pathology that can cause renal or extrarenal manifestations, even leading to chronic kidney disease in patients. These patients can present multiple problems secondary to their diagnosis so sequential surgical treatment can provide alternatives in the final stages of renal pathology.

In the case presented, the patient debuted with symptoms of recurrent urinary tract infection that led to imaging studies that reported the presence of polycystic kidney and liver disease.

Initial imaging studies reported liver and kidneys with lobulated contours, increased dimensions with multiple rounded images, well-defined regular contours of liquid density, and longitudinal measurements of 22.9 cm right and 21.6 cm left.

A complete protocol for ADPKD and chronic kidney disease was initiated as a candidate for donation and sequential surgical procedures were established.

The first procedure consisted of a laparoscopic cholecystectomy plus resection of two giant cysts to reduce probable future risks, which was performed without complications.

Subsequently, the need to perform a bilateral native nephrectomy before or after transplantation was analyzed. The patient was not on renal replacement therapy with hemodialysis or peritoneal dialysis because a previous transplant was desired to allow better graft acceptance, moreover, the native kidneys left space for the graft, so pre-transplant nephrectomy was ruled out.

The renal transplant was successfully performed, presenting immediate function and demonstrating, through a Doppler USG, a renal artery resistance index of 0.5 without the presence of stenosis and thrombosis.

The patient did not present data of recurrent urinary tract infection subsequently, however, bilateral lumbar pain persisted and an imaging study corroborated the decrease in the vital space of the renal graft secondary to the growth of the native kidneys, so it was decided to perform a bilateral native nephrectomy, which did not present complications.

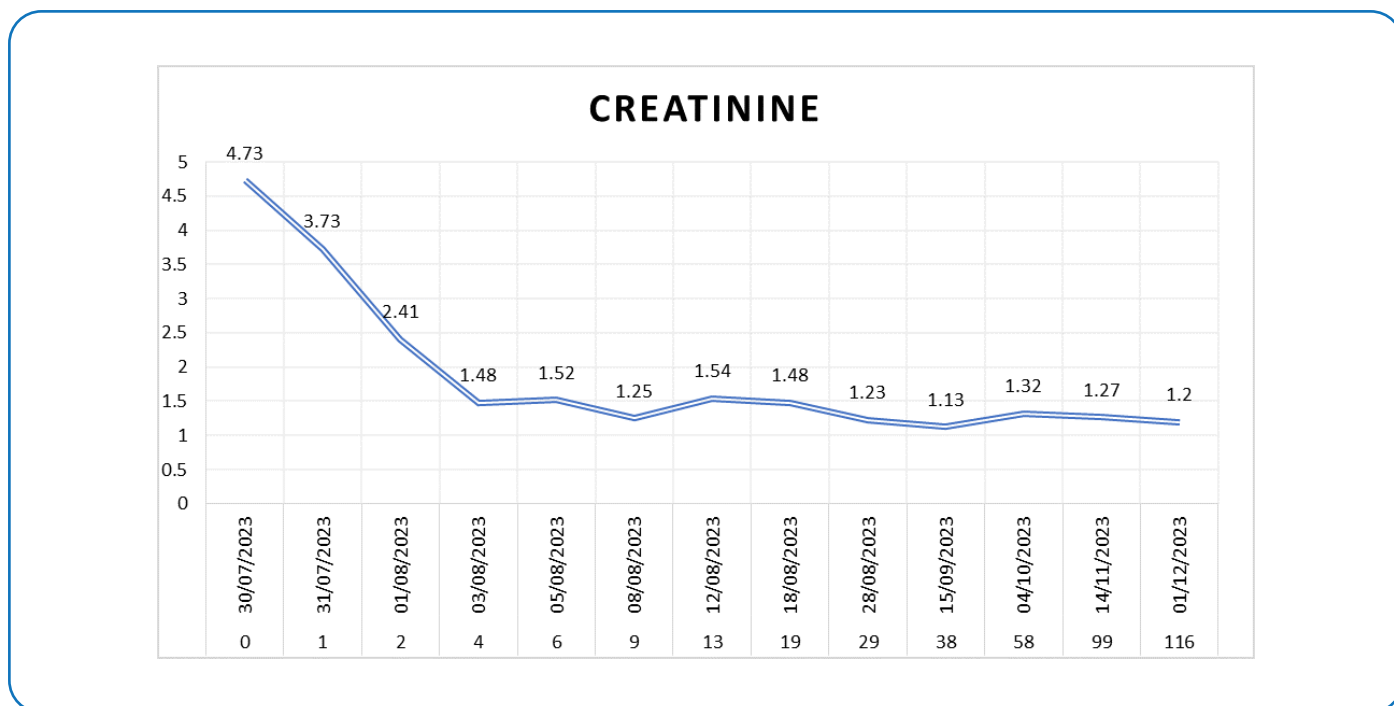
Native nephrectomy is indicated in patients with large kidneys that may cause symptoms of pressure, pain, recurrent infections, bleeding, hypertension, or suspicion of malignancy or to create space for the renal graft [8].

The various literatures mention that in general, patients who receive renal transplantation before nephrectomy do not present a worsening of renal function parameters, and also mention that there is no immunological risk [8].

Regarding the technique of bilateral native nephrectomy, in the patient it was performed by open surgery; in this regard, the literature mentions that there are no inferior results of open surgery compared to laparoscopic surgery. In general, the frequency of complications, intraoperative blood volume loss, or death does not depend on the surgical technique. However, it is mentioned that recovery may be faster with the minimally invasive technique [9].

The sequential surgical treatment was provided to the patient without complications in any of the three interventions, in addition, the support presented throughout the entire protocol was evaluated and in this regard, the literature mentions that the main facilitators of the entire process are social support, including knowledge of the living donor, which in this case was related, financial support and education about the health system [10].

**Table 4:** Pre-transplant creatinine values up to the present day.



Some of the barriers that can occur during treatment include a lack of knowledge of the entire process, receiving insufficient or false information, difficulty in the complete evaluation of the donor or not having social support [10].

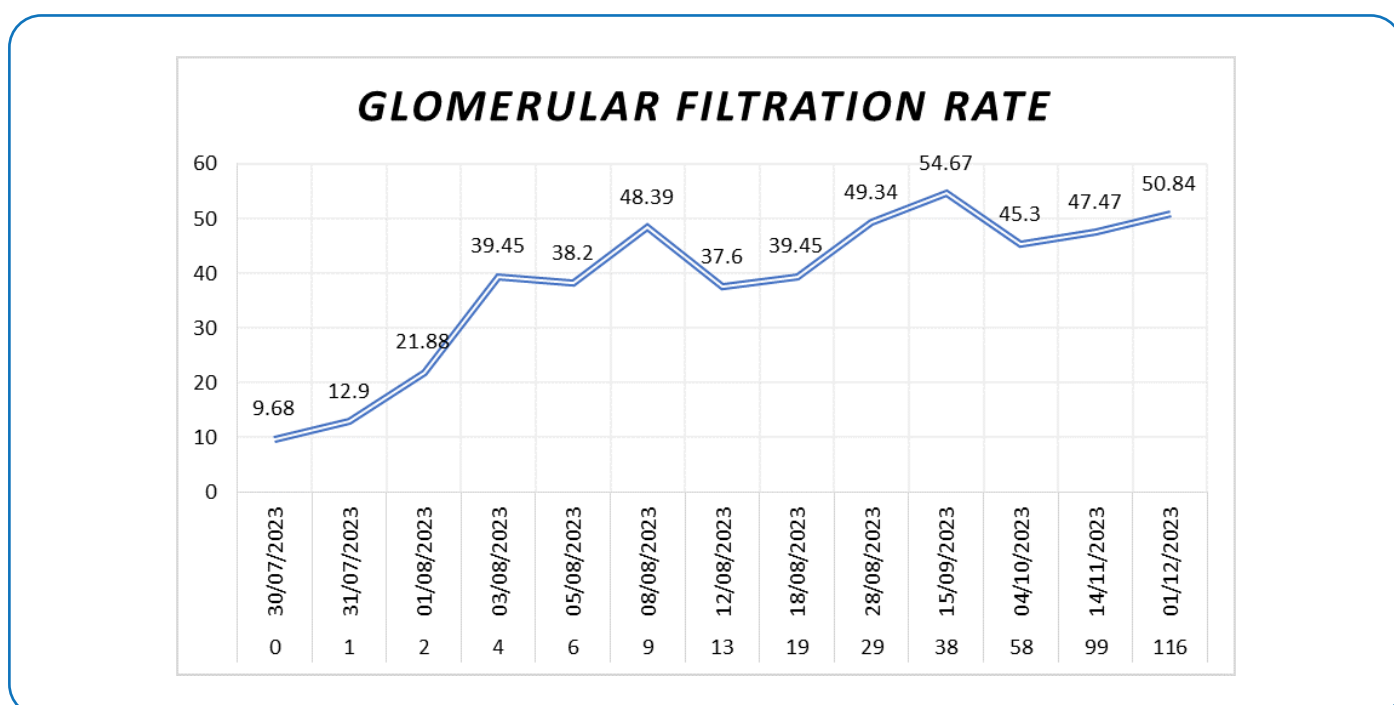
In our case, none of these barriers or impediments were present.

Finally, the prognosis of patients with ADPKD after

transplantation and nephrectomy is good due to a lower risk of recurrence of native disease [5].

In the case of the patient, we followed up with control urinary function tests that report a decrease in creatinine values from 4.73 before renal transplantation to 1.2 mg/dl (Table 4) in her last evaluation and her glomerular filtration rate is currently at 50.84 (Table 5).

**Table 5:** Glomerular filtration rate from pre-transplantation to the present.



## CONCLUSION

In our experience, sequential surgical treatment in patients with ADPKD may provide a safer and recommended approach to achieve adequate renal graft acceptance, including decreasing future surgical risks as well as providing space to decrease the risk of ischemia, thrombosis, or renal vessel stenosis. On the other hand, avoiding future complications due to native kidney permanence such as recurrent urinary tract infections, severe pain, or risk of cancer development.

In the patient, the resection of cysts reduced the possibility of risks in the transplant; in addition, the high specialty surgery with the use of ICG and special sutures reduced ischemia times and provided greater safety to the procedure. Subsequently, a nephrectomy was performed, the indication for which was the risk of graft compromise due to the reduction of the vital space. No complications were observed in the interventions, such as loss of blood volume, immunological alteration, or any graft rejection.

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We recommend providing sequential surgical treatment seeking the social, family, and professional support of patients for a better execution of the protocol, as well as close follow-up of patients during the first post-transplant year and annually thereafter.

Finally, our initial experience can serve as a guide for future studies seeking to generate a sequential surgical treatment of patients with PKD for an adequate acceptance of the graft and improvement in the quality of life.

## CONFLICT OF INTEREST

The authors declare no conflicts of interest.

## CONSENT FOR PUBLICATION

The patient gave written informed consent to the publication of this case report and its accompanying images. On request, a copy of the written consent has been submitted to this Journal.