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ARTICLE **CASE REPORT**

From Cause to Solution of End-stage Renal Disease: Kidney Transplant from a Donor with Polycystic Kidney Disease, Case Report, and Review of the Literature

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INTRODUCTION

Patients with end-stage renal disease (ESRD) have a reduced life expectancy and a worse quality of life compared to the general population. [1] Kidney transplantation is well-established as the treatment of choice for those patients, offering longer survival and better quality of life. [2] Due to the shortage of organs and the rising demand, the use of marginal donors has increased in transplant centers. Despite having lower results than standard criteria, they offer a significant advantage over dialysis. Autosomal dominant polycystic kidney disease (ADPKD) is one of the leading genetic causes of ESRD, marked by the gradual development of bilateral renal cystic changes and subsequent deterioration of renal function. [3] There are reports in the literature of transplants with polycystic kidney donors with normal renal function, with good short- and long-term results.[4] Herein, we report our experience with two successful renal transplants from a young donor with polycystic kidney disease with normal graft function.

CASE PRESENTATION

The donor was a 19-year-old female with an unremarkable past medical and surgical history who presented with multiple injuries, including intracranial hemorrhage, after being involved in a motor vehicle accident. The prognosis was deemed very poor, and the family decided to withdraw care and proceed with organ donation. Her creatinine and BUN at arrival were 0.65 mg/dL and 15 mg/dL, respectively. Standard pre-donation serologic workup was unremarkable, except for positive EBV IgG and negative EBV IgM titers. Pre-donation imaging showed bilateral multiple renal cysts on CT scan, Figures 1. and 2. The donation was pursued as a Donation after Circulatory Death (DCD). Warm Ischemic Time (WIT) was 24 minutes from the extubation to aortic flush with cold Histidine-Tryptophan-Ketoglutarate (HTK), and 16 minutes from agonal breathing to flush. Both kidneys were procured and stored in ice. The left kidney measured 15 cm in length and 7 cm in width, with one artery, one vein, and one ureter, Figure 3. The Right kidney measured 13.5 cm in length, and 7 cm in width, with two arteries,

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one vein, and one ureter (Figure 4). Both kidneys contained multiple cysts consistent with ADPKD. The kidneys were received at our facility in static cold storage after almost 17 hours of cross-clamp due to time spent on allocation and distance from the procurement hospital. Then they were placed on a hypothermic pulsatile perfusion machine with adequate flow and resistance (Table 1).

Figure 1 and 2. Abdominal CT Scan: Enlarged kidneys with bilateral multiple renal cysts

Figure 1

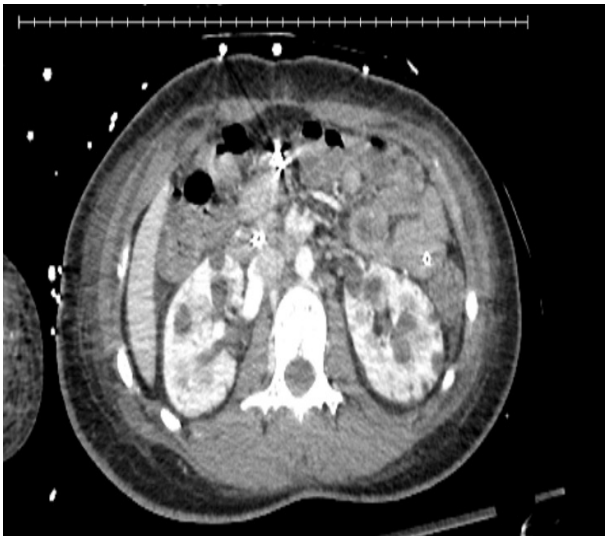


Figure 2

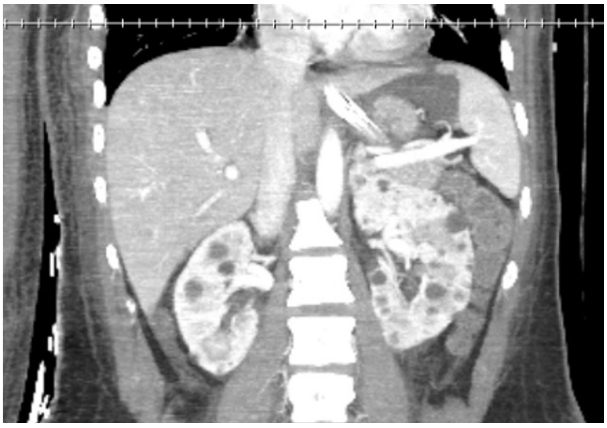


Figure 3 and 4. Anatomy: Left kidney size, 15 x 7 cm, one artery and one vein. Right kidney 13.5 x 7 cm, two artery and 1 vein. Both kidneys with multiple cyst consistent with ADPKD.

Figure 3

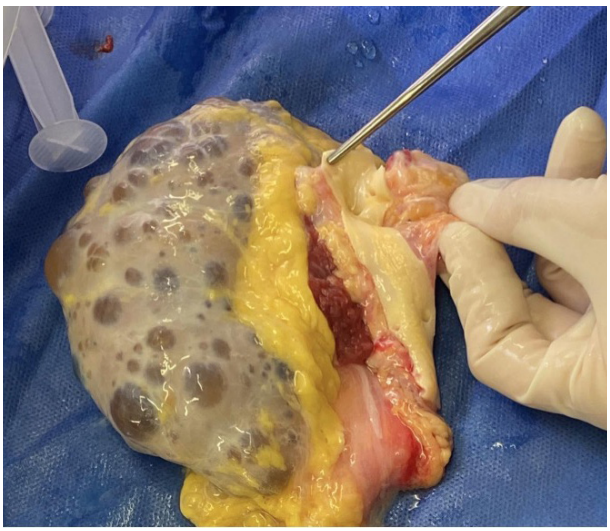


Figure 4

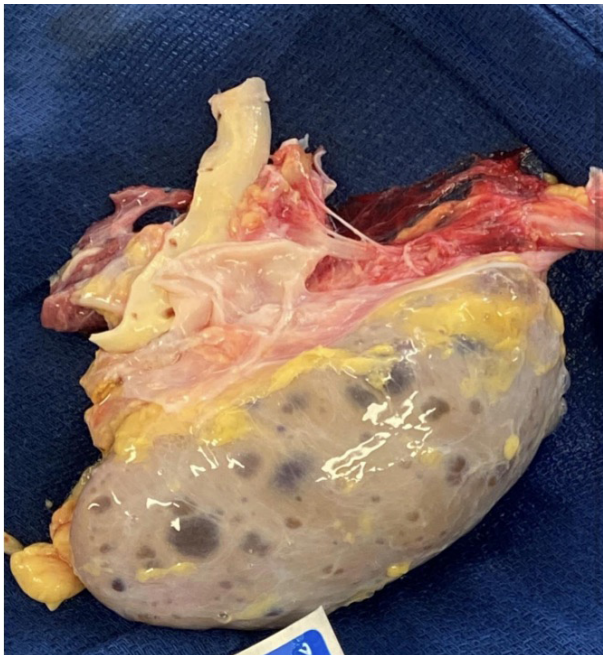


Table 1. Intra-operative renal perfusion data.

	Time	Flow	Resistance	Temperature	Pressure
Right Kidney	0 min	89	0.44	3.1	45/35
	15 min	105	0.32	3.2	40/29
	45 min	167	0.2	3.2	36/34
	75 min	182	0.19	3.1	36/34
	105 min	175	0.18	3.1	35/32

	Time	Flow	Resistance	Temperature	Pressure
Left Kidney	0 min	133	0.19	2	29/22
	30 min	180	0.2	2.1	30/20
	90 min	160	0.13	3.5	25/16
	210 min	162	0.12	3.2	25/15
	270 min	153	0.13	2.8	25/15
	330 min	154	0.12	2.5	25/15
	390 min	170	0.12	2.4	24/20
	410 min	170	0.12	2.3	24/20

RECIPIENT 1- A 68-year-old African American male, BMI 24.71 kg/m², primary diagnosis of ESRD presumably secondary to diabetes millitus – type 2 (DMT2). He has been on hemodialysis support for almost one year. The estimated post-transplant survival score (EPTS) was 65%. He received the right kidney (Figure 4). The WIT was 41 minutes, and the cold ischemic time (CIT) was 19 hours and 13 minutes. The cross-match testing between the donor and recipient 1 was negative. The patient received anti-thymocyte globulin (ATG) induction with methylprednisolone. For maintenance immunosuppression, tacrolimus and mycophenolate mofetil were immediately started postoperatively. The surgical procedure was conducted successfully, without any complications, and immediately restored kidney function (Figures 5 and 6). demonstrate post-op day 1 renal ultrasound. The patient was discharged on post-op day 3, with close outpatient follow-up. At six months post-transplant, the patient continues to have good kidney function (Graphs 1. and 2).

Figure 5 and 6. Post op Day 1 renal ultrasound: Right lower quadrant renal graft measuring approximately 12.6 cm in length with several cysts within the kidney. The renal artery and vein were patent with typical waveforms and no evidence of high-grade stenosis. The main renal artery RI was 0.79, upper pole artery RI of 0.75, mid pole artery RI 0.80, and inferior pole artery RI of 0.77.

Figure 5

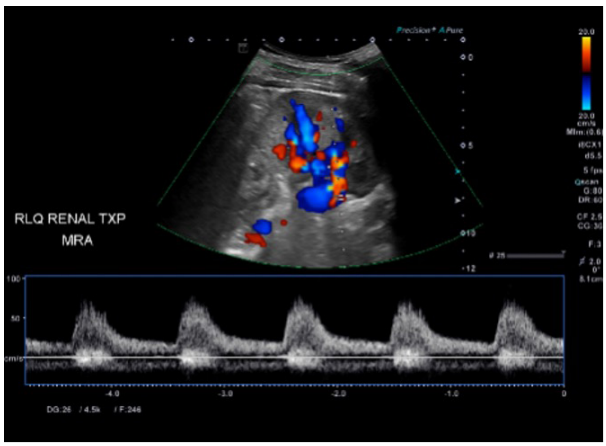
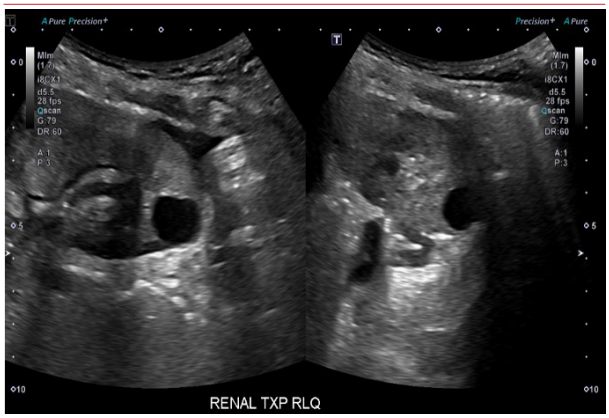


Figure 6



RECIPIENT 2 - A 79-year-old Caucasian female, BMI 29.76 kg/m² with ESRD secondary to chemotherapy (Cytoxan and Daclitaxel) for breast cancer diagnosed ten years previous to the transplant. Her estimated post-transplant survival score (EPTS) was 79%. She received the left kidney (Figure 3). Cross-match testing was negative, WIT was 30 minutes, and CIT of 22 hours and 28 minutes. For induction, she received Basiliximab and methylprednisolone; mycophenolate and Tacrolimus were given immediately postoperatively. The surgical procedure went well without significant concern, and the patient was discharged four days after the surgery. Abdominal ultrasound on post-op day 0 is depicted in Figures 7 and 8. The patient was readmitted five months after the transplant for UTI, which was managed successfully with antibiotics. In that admission, a CT scan and renal US were obtained, and both were unremarkable and showed a stable size of the transplanted kidney compared to pretransplant (Figure 9). At six months post-transplant, the patient continues to have excellent kidney function (Graphs 1 and 2).

Figure 7 and 8. abdominal US day 0: lower quadrant renal transplant measuring approximately 14.7 cm in length. Small volume of free fluid adjacent to the transplanted Kidney, plus multiple simple renal cysts. The renal artery and vein were patent with normal waveforms and no evidence of high-grade stenosis. The main renal artery RI 0.86, upper pole artery RI 0.79, mid pole artery RI 0.79, and inferior pole RI 0.81

Figure 7

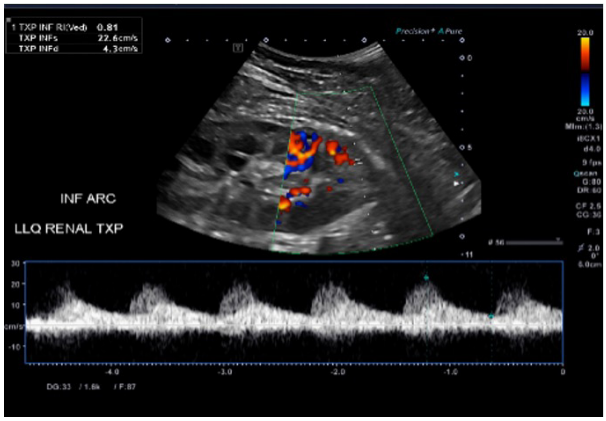


Figure 8

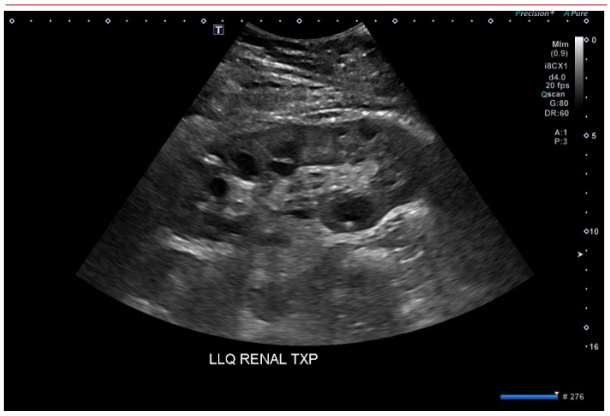
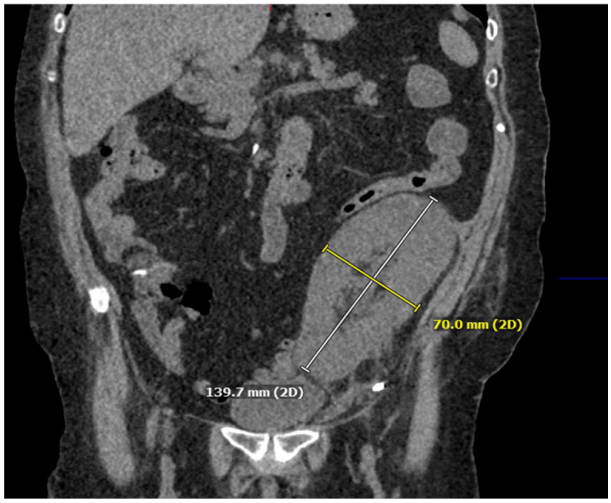
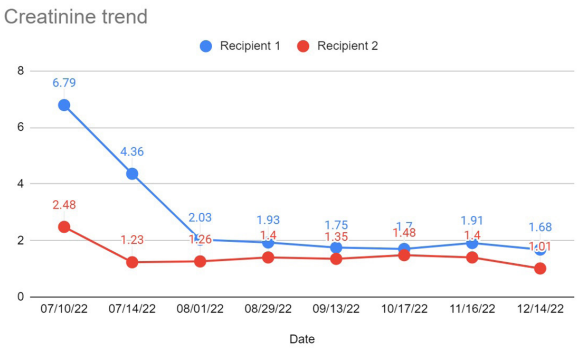


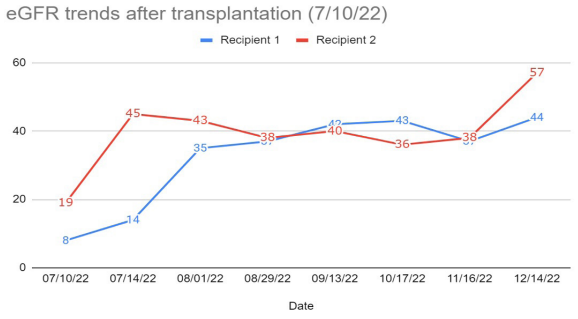
Figure 9. CT abdomen recipient 2, left kidney with stable size 5 months post transplant.



Graph 1. Shows the Creatinine Trend



Graph 2. Shows the eGFR trends after transplantation (7/10/22)



DISCUSSION

For ESRD patients, renal transplant is still the best treatment modality to reduce mortality and improve quality of life. Due to the unmet supply of kidney grafts in high demand worldwide and the increasing number of patients on the waiting list, Several strategies have been implemented to ensure the expansion of the grafts. Although less effective than standard criteria donor (SCD), recipients with marginal kidneys are proven to have better survival than waitlisted patients. Another attempt to deal with the graft shortage is transplanting polycystic kidneys from young donors with normal renal function.

ADPKD is a genetically inherited disorder characterized by the presence of multiple renal cysts and a gradual progression leading to End-Stage Renal Disease (ESRD). It affects all races, with a prevalence of approximately 42.6 per 100,000 persons in the US in recent decades. [6] Mutations in PKD1 and PKD2 genes are the most frequent. Numerous studies estimated a 10-year lag between the onset of symptoms and ESRD. [3;7;8] About 20% of ADPKD patients will develop ESRD at the age of 50 and 60% at 70. [4] Rapid ADPKD progression may be defined as the onset of ESRD at age <55 years. [9]. Several complications related to ADPKD kidneys should be considered including infection, cyst rupture, hemorrhage, and cancer. Renal cell carcinoma (RCC) is found to be more associated with ADPKD; patients with ADPKD and ESRD are 3 times more likely to develop RCC than patients only with ESRD. [13;14] Diagnosing malignancy in polycystic kidney disease is said to be more difficult than with non-cystic kidney disease, as findings suggestive of malignancy may be mistaken for the usual clinical, laboratory, and radiological findings of ADPKD. [15]

There is increased advocacy for using polycystic kidneys for organ donations. Several reports in the literature use polycystic kidneys with promising results. A comprehensive review of all published cases of polycystic donor kidneys between 1980 and 2018 presented 16 different cases of successful polycystic donor kidney transplantation. [4] Issa et al. reported a case where using a polycystic kidney graft, the patient was able to reach an outcome of 20 years of survival. Which is by far the most extended post-transplant graft survival recorded from ADPKD donors. [16] All these advancements encouraged the use of polycystic kidneys for some selected subset of patients, such as older patients on dialysis with a low life expectancy. [17] By using a well-selected polycystic kidney from a young donor for this subset of patients, it is possible to achieve a considerable improvement in their quality of life while shortening the waiting time for transplantation, expanding the donor pool, and, most importantly, attaining a life expectancy after transplant that is comparable to that of kidneys from expanded criteria donors (ECD). [18]

Criteria for the donation of the polycystic kidney have been proposed as follows: length should be

less than 15 cm, donor age less than 50 years, normal creatinine at the time of the procurement, and with an intended CIT of preferably <12 h and not >24 h. [19] The recipient must be fully counseled, and consent must be given. The expected viability of a polycystic kidney graft after transplantation depends mainly on the donor's age, hence the importance of selecting a younger donor to delay the onset of ESRD and avoid increased complications in the recipient. Our donor is a 19-year-old female with a history of polycystic kidney disease but normal kidney function. The patient did not have any previous medical comorbidities. At this age, the patient is far from developing renal failure secondary to her underlined polycystic disease. Using the graft from this patient at this age will probably positively impact the long-term survival of the recipients. The best suitable recipients would be older recipients to avoid long-term complications of polycystic donor kidneys, such as an increase in size, renal dysfunction, and risk of malignancy.

Kidney donor profile index (KDPI) is a method developed to assess donor kidney quality for cadaveric transplants. [20] Grafts with low KDPI scores are expected to perform better than grafts with higher KDPI scores. Putting our patient into the KDPI calculator, we calculated a KDPI score of 11%, which means only 11% of all kidneys would perform better than it. However, since the patient has ADPKD, it is difficult to predict the actual likelihood of performance in the recipient as KDPI does not consider polycystic kidneys in the calculation. We strongly believe that these grafts would yield favorable outcomes, and we encourage using deceased donor kidneys with early ADPKD in selected individuals.

CONCLUSION

Kidneys from selected donors with polycystic kidney disease can be transplanted successfully in selected recipients.

AUTHORS' CONTRIBUTIONS

All authors contributed to the study's conception, design, writing, and review. All authors read and approved the final manuscript.

REFERENCES

1 Turin TC, Tonelli M, Manns BJ, Ravani P, Ahmed SB, Hemmelgarn BR. Chronic kidney disease and life expectancy. Nephrol Dial Transplant. 2012;27(8):3182-6.

2 Wolfe RA, Ashby VB, Milford EL, Ojo AO, Ettenger RE, Agodoa LY, et al. Comparison of mortality in all patients on dialysis, patients on dialysis awaiting transplantation, and recipients of a first cadaveric transplant. N Engl J Med. 1999;341(23):1725-30.

3 Woon C, Bielinski-Bradbury A, O'Reilly K, Robinson P. A systematic review of the predictors of disease progression in patients with autosomal dominant polycystic kidney disease. BMC Nephrol. 2015;16:140.

4 Shamali A, Milsom-Mcquillan S, Gibbs P. Outcomes of renal transplant from donors with polycystic kidney disease. Int J Surg. 2018;51:229-32.

5 Abouna GM. Organ shortage crisis: problems and possible solutions. Transplant Proc. 2008;40(1):34-8.

6 Aung TT, Bhandari SK, Chen Q, Malik FT, Willey CJ, Reynolds K, Jacobsen SJ, Sim JJ. Autosomal Dominant Polycystic Kidney Disease Prevalence among a Racially Diverse United States Population, 2002 through 2018. Kidney360. 2021 Sep 22;2(12):2010-2015. doi: 10.34067/KID.0004522021. PMID: 35419536; PMCID: PMC8986058.

7 Churchill DN, Bear JC, Morgan J, Payne RH, McManamon PJ, Gault MH. Prognosis of adult onset polycystic kidney disease re-evaluated. Kidney Int. 1984;26(2):190-3.

8 Parfrey PS, Bear JC, Morgan J, Cramer BC, McManamon PJ, Gault MH, et al. The diagnosis and prognosis of autosomal dominant polycystic kidney disease. N Engl J Med. 1990;323(16):1085-90.

9 Schrier RW, Brosnahan G, Cadnapaphornchai MA, Chonchol M, Friend K, Gitomer B, et al. Predictors of autosomal dominant polycystic kidney disease progression. J Am Soc Nephrol. 2014;25(11):2399-418.

10 Cachat, Francois; Renella, Raffaele (2016). Risk of cancer in patients with polycystic kidney disease. The Lancet Oncology, 17(11), e474–. doi:10.1016/S1470-2045(16)30529-0

11 Yu TM, Chuang YW, Yu MC, et al. Risk of cancer in patients with polycystic kidney disease: a propensity-score matched analysis of a nationwide, population-based cohort study. Lancet Oncol 2016; 17: 1419–25

12 Altheaby A, Almukhlifi A, Aldoukhi A, Alfaleh A, Aboalsamah G, Alshareef A, et al. Why Living Kidney Donor Candidates Are Turned Down? A Single-Center Cohort Study. Cureus. 2020;12(8):e9877.

13 Hajj P, Ferlicot S, Massoud W, Awad A, Hammoudi Y, Charpentier B, et al. Prevalence of renal cell carcinoma in patients with autosomal dominant polycystic kidney disease and chronic renal failure. Urology. 2009;74(3):631-4.

14 Jilg CA, Drendel V, Bacher J, Pisarski P, Neeff H, Drognitz O, et al. Autosomal dominant polycystic kidney disease: prevalence of renal neoplasias in surgical kidney specimens. Nephron Clin Pract. 2013;123(1-2):13-21.

15 Gomez G, Althaus A, Gruessner CE, Hirsch MS, Steele GS. Clear cell tubopapillary renal cell carcinoma mimicking polycystic kidney disease: A case report. Urol Case Rep. 2018;16:35-7.

16 Issa N, Chedid M, Irazabal MV, Dean PG, Chebib FT. Twenty-Year Survival of Kidney Transplant From a Deceased Donor With Autosomal Dominant Polycystic Kidney Disease. Kidney Int Rep. 2021;6(8):2240-2.

17 Ojo AO. Expanded criteria donors: process and outcomes. Semin Dial. 2005;18(6):463-8.

18 Bagul A, Olsburgh JD, Calder F. Deceased Polycystic Kidney Donors - Should Be Considered to Expand the Donor Pool: 2480. Transplantation. 2012;94(10S):606-7.

19 Olsburgh JD, Godbole HC, O'Donnell PJ, Koffman GC, Taylor JD, Khan MS. Transplantation of kidneys from deceased adult polycystic donors. Am J Transplant. 2006;6(11):2809-11.

20 Baloglu I, Tonbul HZ, Turkmen K, Selcuk NY, Iyisoy MS. Are Kidney Donor Risk Index/Kidney Donor Profile Index Scores Predictor of Future Graft Function? Saudi J Kidney Dis Transpl. 2021;32(4):979-85.