











Nephrectomy in autosomal dominant polycystic kidney disease: a consensus statement of the ERA Genes & Kidney Working Group

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ABSTRACT

A substantial number of patients with autosomal dominant polycystic kidney disease (ADPKD) undergo a nephrectomy, especially in workup for a kidney transplantation. Currently, there is no evidence-based algorithm to guide clinicians about which patients should undergo nephrectomy, the optimal timing of this procedure, or the preferred surgical technique. This systematic review-based

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consensus statement aimed to answer important questions regarding nephrectomy in ADPKD. A literature review was performed and extended to a meta-analysis when possible. For this purpose, PubMed and EMBASE were searched up to May 2024. Fifty-four publications, describing a total of 2391 procedures, were included. In addition, an exploratory questionnaire was sent to urologists, nephrologists, and transplant surgeons. These sources were used to develop practice points about indications, complications, mortality, and timing and technique of nephrectomy. In addition, data on renal embolization as a potential alternative to nephrectomy were explored and summarized. To reach consensus, practice points were defined and improved in three Delphi survey rounds by experts of the European Renal Association Working Group Genes & Kidney and the European Association of Urology Section of Transplantation Urology. A total of 23 practice points/statements were developed, all of which reached consensus. Among others, it was deemed that nephrectomy can be performed successfully for various indications and is an intermediate risk procedure with acceptable mortality and minimal impact on kidney graft function when performed before, in the same session or after transplantation. The complication rate seems to increase when the procedure is performed as an emergency. During the workup for transplantation, patient complaints should be assessed routinely by questionnaires to indicate symptom burden. Deciding on the need for nephrectomy and exploring potential alternatives such as kidney embolization should be a process of shared decision-making, preferably after multidisciplinary consultation.

Keywords: ADPKD, kidney transplantation, nephrectomy, polycystic kidney disease

INTRODUCTION

Autosomal dominant polycystic kidney disease (ADPKD) is the most common hereditary kidney disorder. It is characterized by the formation and growth of numerous cysts in both kidneys, which compress viable kidney tissue, leading to a decline in kidney function. Ultimately, the majority of affected patients require kidney replacement therapy, typically at a median age around 58 years [1].

When patients with ADPKD reach kidney failure, kidney transplantation is the preferred modality in most patients [2, 3]. In a substantial number of patients, nephrectomy of one or both native kidneys is performed in the time period around a transplantation [4, 5]. Indications for nephrectomy include, but are not limited to, lack of space for a kidney allograft, uncontrolled refractory pain, cyst infection, persistent cyst hemorrhage, urolithiasis, or malignancy [6–8]. How often these procedures are performed varies between hospitals, ranging from 0% to 90% of patients with ADPKD who are either planned for or have received a kidney transplantation [8, 9]. Regarding the timing, nephrectomy can be performed pre-transplantation, combined with transplantation, or post-transplantation [5, 6, 10–12]. The most commonly employed surgical techniques include open and (hand-assisted) laparoscopic approaches, either uni- or bilaterally [4, 9, 13–15].

At present, there is no evidence-based algorithm to guide clinicians on which patients with ADPKD should undergo nephrectomy, the optimal timing of the procedure, or the preferred technique. Protocols vary among hospitals, highlighting the lack of consensus on the approach to nephrectomy in ADPKD. To address this knowledge gap, an expert team in the University Medical Center of Groningen, the Netherlands, was asked by the European Renal Association (ERA) Working Group Genes & Kidney to conduct a comprehensive systematic literature review, and to summarize the results in a meta-analysis whenever feasible. Additionally, a questionnaire was distributed to nephrologists, urologists, and transplantation surgeons. The results from these information sources were utilized by the expert team, the board of the ERA Working Group Genes & Kidney, the board of the European Association of Urology (EAU) Section of Transplantation Urology, patient representatives, and external experts to address clinically significant questions regarding nephrectomies in patients with ADPKD. These questions included determining the indications for nephrectomy, identifying the operative techniques used for this procedure, assessing potential complications, and determining the optimal timing for nephrectomy in relation to kidney transplantation. Besides, currently available data on renal emboliza-

tion as a potential alternative to nephrectomy were explored and summarized. This resulted in the development of practice points, for which consensus was sought through a Delphi survey.

Recently, KDIGO issued a nephrology guideline on the diagnosis and treatment of ADPKD [16]. This guideline also mentions some considerations about when to perform a nephrectomy. The present manuscript endorses this work and extends it by giving a comprehensive overview of more aspects, including a multidisciplinary view, and also covers surgical and radiological aspects.

METHODS

Systematic literature review

Data sources and searches

An electronic literature search was conducted on 7 May 2024. PubMed and EMBASE were queried for articles published since January 1950 using the search string: ['polycystic kidney disease' OR 'ADPKD' AND 'nephrectomy'], which yielded a total of 2942 publications. Reference lists of retrieved articles were manually reviewed to identify any additional relevant publications. After excluding 559 duplicates and 33 publications not published in English, two investigators (P.G. and N.F.C.) independently assessed the titles and abstracts of the remaining articles for relevance. Any disagreements between the investigators were resolved through discussion. Figure 1 provides an overview of the literature search process. This systematic review adheres to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines (Supplementary Table S1) [17].

Study selection

Three hundred and twenty-six publications were retrieved for full text evaluation. Two hundred and seventy-four publications were excluded for the following reasons: full text not available ($n = 148$), less than 10 nephrectomy procedures described ($n = 79$), review or meta-analysis ($n = 5$), no (original) nephrectomy data ($n = 33$), nephrectomy performed together with embolization ($n = 5$), inclusion of non-ADPKD patients ($n = 2$), or indications for nephrectomy not given per procedure ($n = 2$). As a result, 54 articles were included in this review, listed in Supplementary Table S2. Of these, 49 were used to assess indications for nephrectomy, 12 to evaluate the timing of nephrectomy, 13 to examine differences between operative techniques, and 47 to assess complications. In total, the included publications describe 2391 nephrectomy procedures.

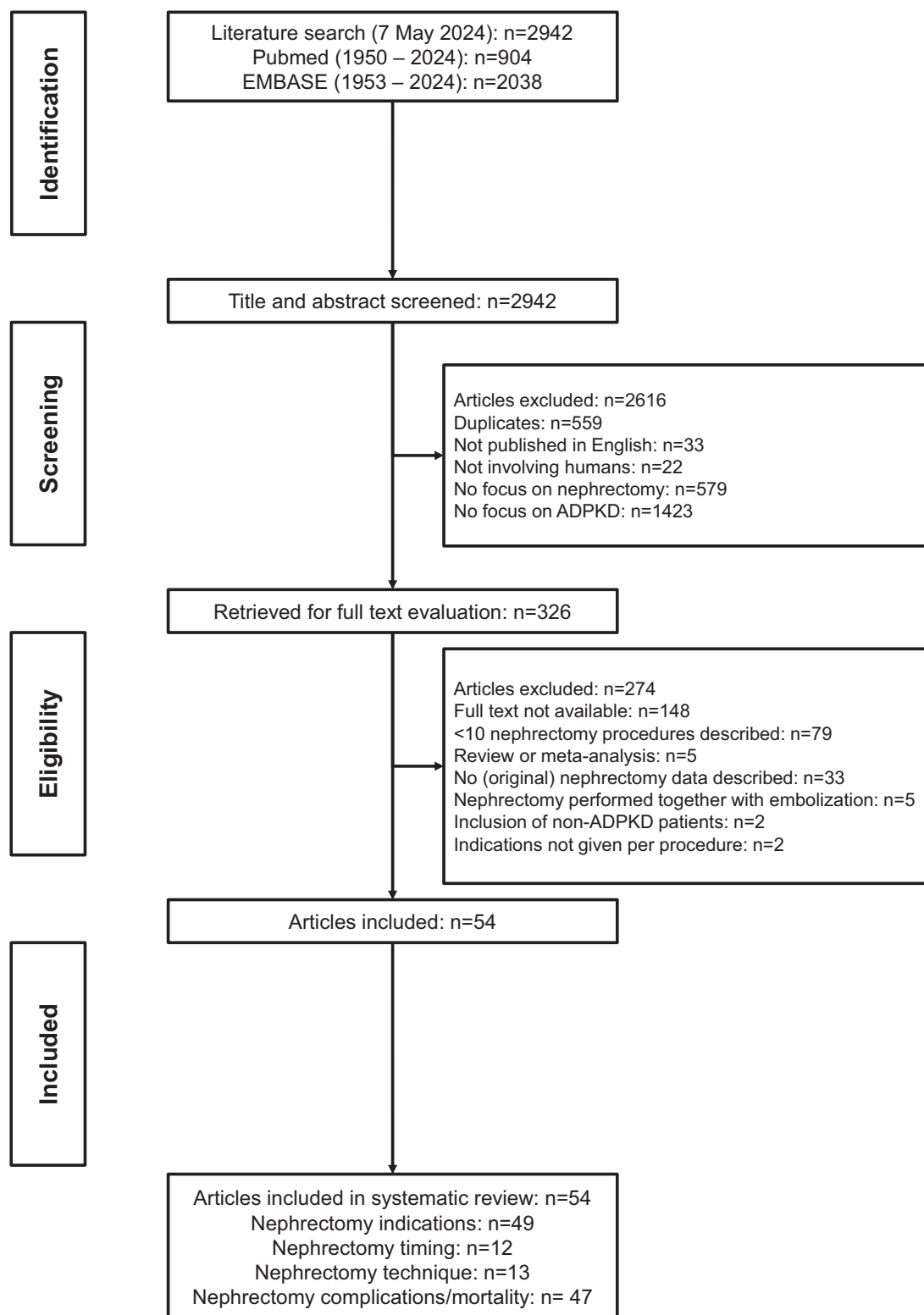


Figure 1: Flowchart of literature search.

Data extraction and data analysis

When analyzing indications for nephrectomy, several indications could be assigned to a single procedure. The variables used to assess the operative technique, timing related to kidney transplantation, and complications included age, sex, operative time (minutes), hospital stay (days), number of blood transfusions per pro-

cedure, number of major peri-/postoperative complications per patient, kidney weight (grams), and donor type (deceased/living). Complications were rated according to the Clavien–Dindo classification [18]. Major complications were defined as Clavien–Dindo classification ≥ 3 . For these variables, means and standard deviation or frequencies were collected from the identified

publications. If a mean and *P*-value were available, a standard deviation was calculated using the Revman calculator of the Cochrane Library [19]. When only the median and (interquartile) range were available, a mean and standard deviation were calculated using a method described by Wan et al. [20]. In case no information concerning a specific variable was available, we contacted the corresponding and/or lead author to obtain the necessary information.

Assessing risk of bias

Risk of bias was assessed using the Newcastle–Ottawa Scale [21]. Two investigators independently assessed the publications. Each study could receive a maximum score of nine stars. Publications with zero to three stars were categorized as having a high risk of bias, those with four to six stars as having an intermediate risk of bias, and those with seven to nine stars as having a low risk of bias. Out of 23 publications, 22 were found to have a low risk of bias (eight stars: *n* = 18, seven stars: *n* = 4), and one publication was categorized as having an intermediate risk of bias (six stars) (Supplementary Table S3).

Statistical analysis

When data allowed, a formal meta-analysis was performed, choosing a random effects model to account for heterogeneity. Heterogeneity was assessed using the *I*² statistic. An *I*² > 50% was considered to indicate substantial heterogeneity. Differences in continuous outcomes are described as mean difference (MD) [95% confidence interval (CI)] and differences in incidence as risk difference (RD) [95% CI]. Meta-analyses were visualized using forest plots. Normally distributed continuous data and non-normally distributed continuous data were expressed as mean ± standard deviation (SD) or median (interquartile range), respectively. Nominal data were presented as percentages. Statistical significance was defined as a two-sided *P*-value of <0.05. Sensitivity analyses were conducted to identify outliers with a 95% CI outside of the 95% CI of the pooled effect. The leave-one-out method was used to assess effect sizes and *I*² values after each study was excluded. Due to the nature of the included studies (case series), no assessment of reporting bias was performed. All statistical analyses and data visualization were performed using R version 4.0.5 (Vienna, Austria).

Exploratory questionnaire

To explore the current clinical practice concerning native nephrectomy in patients with ADPKD, a digital questionnaire comprising 30 questions was developed by a multidisciplinary team at the University Medical Center Groningen, the Netherlands, including nephrologists, urologists, transplant surgeons, and patient representatives. The questionnaire covered topics such as physician demographics, indications for nephrectomy, surgical procedures and techniques, kidney embolization, and the postoperative period. Ninety-eight physicians were invited to participate. These physicians included nephrologists (*n* = 45), urologists (*n* = 32), and transplant surgeons (*n* = 21). Invitations, along with an explanatory letter, were sent by email. A reminder was sent after 1 month, followed by a second reminder after 2 months. In total, 56 physicians (57%) responded to the questionnaire (Supplementary Table S4). Their mean age was 49 ± 7 years, and 66.1% were male. The majority (69.6%) worked in a tertiary medical center and had a median of 14 [9–22] years of professional experience. These physicians consulted a median of 20 (10–30) patients with ADPKD annually, with

8 [3–15] of them undergoing evaluation for kidney transplantation. On average, these physicians reported 3 [2–5] nephrectomies performed in patients with ADPKD at their hospital per year.

Delphi survey

Practice points were created using results of the literature search, meta-analysis, and questionnaire. These practice points were then presented to members of the ERA Working Group Genes & Kidney in all rounds of the survey as well as external ADPKD experts, including nephrologists, interventional radiologists, urologists, and transplant surgeons, and the EAU Section of Transplantation Urology in all subsequent rounds after the first round of the Delphi survey. Possible options in the Delphi survey were 'Fully agree', 'Partially agree', 'Partially disagree', 'Disagree', and 'I abstain'. The consensus threshold for acceptance of a practice point was reached when ≥70% of respondents fully agreed and ≥90% fully or partially agreed. After the first round of the Delphi survey, an online consensus meeting was held to discuss the practice points that did not reach consensus and practice points that did but received major comments. Respondents could choose to provide contact information to be invited for future rounds of the survey, in which improved practice points were evaluated. Ultimately, three rounds were needed to reach consensus on all points. Respondent characteristics and results of the Delphi survey are depicted in Supplementary Tables S5 and S6. The numbers of respondents in the first, second and third rounds of the Delphi survey were 56, 60, and 52, respectively. The percentages of previous respondents who completed the second and third surveys were 100 and 89.8, respectively. Respondents of the Delphi surveys were employed in a total of 21 countries, with the most respondents working in Germany (11.7%), Belgium (10.0%), Spain (10.0%), the Netherlands (8.3%), the USA (8.3%), France (6.7%), and Japan (6.7%). Most respondents were nephrologists (78.3%) and urologists (16.7%). The majority of respondents had >15 years of work experience and 88.3% of respondents only saw adult patients with ADPKD. Per year, 36.7% saw 11–50, 25.0% 51–100, and 28.3% >100 patients with ADPKD. The majority of respondents were involved in the decision process around nephrectomy in patients with ADPKD in 1–5 (46.7%) or 6–20 (41.7%) procedures per year.

RESULTS

What are potential indications for nephrectomy in patients with ADPKD?

For the analysis of nephrectomy indications, we included 49 publications describing a total of 1955 procedures (Table 1). These publications revealed that nephrectomy in patients with ADPKD was performed for various indications, including: lack of space for a kidney allograft (42.6% [95% CI 40.4–44.8]), pain (31.3% [95% CI 29.2 to 33.4]), cyst/urinary tract infections (22.9% [95% CI 21.0–24.7]), cyst hemorrhage/hematuria (14.4% [95% CI 12.8–15.9]), gastrointestinal complaints (7.9% [95% CI 6.7–9.1]), dyspnea (2.4% [95% CI 1.7–3.0]), kidney stones (2.0% [95% CI 1.4–2.6]), and malignancy (1.7% [95% CI 1.2–2.3]). When comparing publications focused on nephrectomy in the transplantation period (*n* = 33; 1497 procedures) with publications describing nephrectomy not specifically related to transplantation (*n* = 16; 458 procedures), differences in indications for nephrectomy were observed. For instance, indications such as lack of space for a kidney allograft (46.6% versus 29.5%, respectively), cyst/urinary tract infections (19.8% versus 33.0%), gastrointestinal complaints (9.2% versus 3.7%), and

Table 1: Indications for nephrectomy in ADPKD.

Publication	Nephrectomy procedures (n)	Lack of space for kidney allograft, n (%)	Pain, n (%)	Cyst/urinary tract infections, n (%)	Cyst hemorrhage/hematuria, n (%)	Gastrointestinal complaints, n (%)	Dyspnea, n (%)	Kidney stones, n (%)	Malignancy, n (%)	Other, n (%)
Publications focused on nephrectomy performed during the period surrounding transplantation										
Bennett et al., 1973 [47]	31	16 (52)		11 (35)	12 (39)					
Hadimeri et al., 1997 [41]	26	12 (46)	3 (12)	5 (19)	3 (12)				3 (12)	4 (15)
Glassman et al., 2000 [74]	10	7 (70)	1 (10)	1 (10)	2 (20)	1 (10)				
Fuller et al., 2005 [75]	32	8 (25)	8 (25)	8 (25)	4 (13)	2 (6)			2 (6)	
Rozanski et al., 2005 [96]	30	27 (90)		1 (3)	1 (3)			1 (3)		
Ismail et al., 2005 [51]	16	12 (75)		1 (6)	3 (19)					
Tabibi et al., 2005 [22]	13	13 (100)								
Nunes et al., 2007 [23]	16	16 (100)								
Wagner et al., 2007 [48]	32	31 (97)	11 (34)	9 (28)	5 (16)	31 (97)				
Kramer et al., 2009 [25]	20		18 (90)	5 (25)		1 (5)	2 (10)	1 (5)		
Lucas et al., 2010 [9]	42		35 (83)	8 (19)		16 (38)	5 (12)			
Kirkman et al., 2011 [4]	35	14 (40)	26 (74)	5 (14)	6 (17)	4 (11)				
Patel et al., 2011 [26]	31	1 (3)	12 (39)	14 (45)	1 (3)				3 (10)	
Skauby et al., 2012 [49]	78	43 (55)	17 (22)	16 (21)	19 (24)					4 (5)
Eng et al., 2013 [13]	76	58 (76)	16 (21)	2 (3)	7 (9)				2 (3)	
Chebib et al., 2015 [5]	114	9 (8)	74 (65)	16 (14)	27 (24)	9 (8)	5 (4)		8 (7)	
García-Rubio et al., 2015 [82]	27	20 (74)	2 (7)		5 (19)					
Kim et al., 2016 [40]	22	10 (46)	4 (18)	5 (23)	4 (18)					
Veroux et al., 2016 [94]	65	60 (92)		1 (2)	3 (4)					
Ahmad et al., 2016 [10]	72	25 (35)	55 (76)	15 (21)	17 (24)	26 (36)	12 (17)	5 (7)		8 (11)
Anselmo et al., 2019 [77]	53	34 (64)		11 (21)	7 (13)					
Ietto et al., 2019 [50]	33	14 (42)	2 (6)	10 (30)	7 (21)					
Maxeiner et al., 2019 [27]	121		64 (52)	37 (31)	6 (5)	2 (2)	18 (35)	12 (10)		6 (12)
Abrol and Prieto, 2020 [89]	51		26 (51)	9 (18)	7 (13.7)	18 (35)				
Rosenberg et al., 2021 [30]	12	10 (83)	1 (8)	1 (8)	2 (16.7)					
Mansbridge et al., 2021 [54]	32	9 (28)		5 (16)	1 (3)	10 (31)			5 (16)	2 (6)
Casteleijn et al., 2022 [6]	172	67 (39)	36 (21)	68 (40)	34 (19.8)	15 (9)		1 (1)		
Darius et al., 2022 [8]	77	74 (96)	29 (38)	11 (14)	30 (39.0)	3 (4)		9 (12)		
Rasmussen et al., 2022 [32]	28	26 (93)	1 (4)						1 (4)	
Huynh et al., 2022 [79]	35	23 (66)	19 (54)	5 (14)	7 (20.0)					
Thornas et al., 2023 [86]	24	7 (29)	11 (46)	4 (17)	2 (8)					
Masterson et al., 2023 [92]	14	9 (64)	7 (50)	1 (7)	2 (14)					
Lyu et al., 2024 [81]	57	43 (75)		11 (19)	5 (9)					
Overall nephrectomy related to transplantation	1497	46.6%	31.9%	19.8%	15.3%	9.2%	2.8%	1.6%	1.9%	1.6%

Table 1: Continued

Publication	Nephrectomy procedures (n)	Lack of space for kidney allograft, n (%)	Pain, n (%)	Cyst/urinary tract infections, n (%)	Cyst hemorrhage/hematuria, n (%)	Gastrointestinal complaints, n (%)	Dyspnea, n (%)	Kidney stones, n (%)	Malignancy, n (%)	Other, n (%)
Publications describing nephrectomy not specifically related to transplantation										
Zeier et al., 1992 [37]	47			30 (64)	7 (15)			8 (17)		2 (4)
Seshadri et al., 2001 [87]	20	11 (55)	7 (35)	4 (20)	1 (5)					
Gill et al., 2001 [7]	20	8 (40)	8 (40)	3 (15)	4 (20)				1 (5)	
Bendavid et al., 2004 [52]	22	6 (27)	12 (55)	2 (9)	3 (14)	5 (25)	4 (20)			
Whitten et al., 2006 [84]	10	2 (20)	8 (80)							
Binsaleh et al., 2006 [53]**	6	3 (50)	2 (33)		1 (17)					
Lipke et al., 2007 [88]	18		16 (89)	2 (11)						
Binsaleh et al., 2008 [12]	16	8 (50)	9 (56)	1 (6)	1 (6)					
Desai PJ et al., 2008 [11]	12	1 (8)	8 (67)	2 (17)		2 (17)				
Verhoest et al., 2012 [15]	40	32 (80)	5 (13)		1 (3)	3 (8)		2 (5)		7 (18)
Martin et al., 2012 [99]*	15	4 (27)	13 (87)	2 (13)	1 (7)	7 (47)				
Bansal et al., 2014 [98]	39	21 (54)	16 (41)	2 (5)	2 (5)				1 (3)	
Asimakopoulos et al., 2015 [24]	19	19 (100)	4 (21)		1 (5)					1 (5)
Chen et al., 2018 [100]	33			4 (12)	21 (64)				8 (24)	
Bellini et al., 2019 [14]	33	20 (61)	8 (24)	12 (36)	6 (18)					
Lubennikov et al., 2021 [38]	108		18 (17)	87 (81)	3 (2.8)					
Overall nephrectomy not related to transplantation	458	29.7%	29.3%	33.0%	11.4%	3.7%	0.9%	2.2%	2.2%	2.2%
Overall total publications	1955	42.6%	31.3%	22.9%	14.4%	7.9%	2.4%	1.7%	2.0%	1.7%

*Exact indications of 15 out of 37 nephrectomies given.

**Indications of 6 laparoscopic procedures are used in indications of Binsaleh et al., 2008.

dyspnea (2.8% versus 0.9%) varied. In the questionnaire, physicians estimated that 61.2% of procedures were performed due to lack of space for a kidney allograft, 17.0% for cyst/urinary tract infections, 7.8% for pain, 6.4% for cyst hemorrhage/hematuria, 2.8% for malignancy, 2.5% for gastrointestinal complaints, 0.7% for kidney stones, and 0.5% for a disturbed self-image caused by ADPKD-related abdominal volume growth (Supplementary Table S4).

The percentage of nephrectomies performed to create sufficient space for a kidney allograft varies widely among publications. As expected, papers focused on nephrectomy performed during the transplantation period report a higher percentage of nephrectomies performed for this indication than those not specifically related to transplantation. In some studies, all nephrectomies were performed due to lack of space [22–24]. However, others described limited numbers for this indication [5, 9, 25–28]. Chebib *et al.*, for instance, reported that in their high-volume hospital, lack of space has not been present as an indication for nephrectomy since 2004. In all transplanted patients described in this publication, there was enough space for a kidney allograft [5]. Nonetheless, even in publications where nephrectomies are reported to be performed for the indication lack of space, the criteria for deciding if the polycystic kidney is not excessively enlarged to allow successful kidney allograft placement remain unclear. Some authors suggest markers such as total kidney volume assessed by magnetic resonance imaging (MRI) or kidney length assessed by ultrasound scanning as criteria, but these markers do not account for other abdominal volume-increasing processes, such as an enlarged polycystic liver or patient height [29–31]. Other publications mention that nephrectomy is indicated to create space for a kidney allograft when the native polycystic kidney extends below the iliac crest [30, 32, 33]. As no precise guidelines exist, the decision to remove a native kidney to create space for an allograft or to alleviate complaints related to a high intra-abdominal volume remains surgeon-dependent.

As described earlier, nephrectomy can also relieve ADPKD-related pain complaints. Data from a treatment protocol for ADPKD-related pain, published in 2017 and 2022, demonstrated that nephrectomy effectively relieved intractable chronic pain in patients with ADPKD [34, 35]. However, it should be noted that this procedure is considered a last resort option in patients not on kidney function replacement therapy as the procedure will shorten the time to dialysis and/or transplantation [35]. Furthermore, in a publication from 2023 it was found that nephrectomy positively affected gastrointestinal complaints in patients with ADPKD [36]. To our knowledge, there are no publications investigating the effect of nephrectomy on dyspnea and patient self-image and this should thus be investigated in future studies.

To summarize, the most frequent indication for nephrectomy is lack of space for a kidney allograft, followed by pain, cyst/urinary tract infections, cyst hemorrhage/hematuria, and gastrointestinal complaints. Since these data can only be extracted from patient files, it may be important to adequately record the nephrectomy indication in patient files. It is advised that the possibility of nephrectomy should be mentioned to all patients in the workup for transplantation. Patients will often wonder whether it is possible to remove a kidney, even when this is deemed unnecessary by their physician. A retrospective study published in 2023, investigated quality of life in transplant patients who did and did not undergo a nephrectomy and found a similar quality of life at long-term follow-up [36]. Notably, although quality of life at long-term follow-up was similar be-

tween these groups, 20% of patients who had not undergone nephrectomy indicated that they did wish to have undergone a nephrectomy in hindsight and 96% of patients indicated that the decision not to perform nephrectomy was made by their treating physician, underlining the need to inform patients about the procedure [36]. Mentioning the possibility of nephrectomy to all patients serves as an opportunity to discuss their complaints and potential wish for a nephrectomy. When there are no indications for nephrectomy, the patient can be reassured that nephrectomy is not needed and potential doubts concerning this topic can be alleviated. After a multidisciplinary evaluation during the workup for transplantation by a nephrologist, transplant surgeon, and urologist, the benefits and risks of a nephrectomy can be discussed with the patient, after which a shared decision can be made. A visual summary of practice points concerning general suggestions and indications has been provided in Fig. 2.

Statement 1: There are several potential indications to perform a nephrectomy of native polycystic kidneys, especially during the workup for a kidney transplantation, for instance recurrent cyst infections, recurrent cyst hemorrhage, intractable chronic pain, severe high abdominal volume-related gastrointestinal complaints, recurrent kidney stones, suspected malignancy, and insufficient space to allow a kidney transplantation.

Practice point 1.1: To address questions patients might have, now or in the future, the possibility of a nephrectomy of one or both native polycystic kidneys should be mentioned to all patients with ADPKD during the workup for a kidney transplantation.

Practice point 1.2: Deciding which patients will have a nephrectomy of one or both native polycystic kidneys should be a process of shared decision-making, preferably after multidisciplinary consultation.

Practice point 2: Nephrectomy can be an effective treatment for intractable chronic ADPKD-related pain and severe gastrointestinal complaints, with a lower threshold in the post-transplantation period and a high threshold before kidney failure has occurred, due to its impact on kidney function.

When should nephrectomy be performed in cases of recurrent cyst infections?

While many publications have discussed nephrectomy for recurrent cyst infections [26, 37–41], no data define the exact number or severity of cyst infections warranting a nephrectomy. In our questionnaire, 60.7% of physicians deemed two to three cyst infections in 2 years sufficient to merit a pre-transplantation nephrectomy, while 26.8% indicated that nephrectomy might be indicated after four to five cyst infections in 2 years pre-transplantation. Therefore, during the pre-transplantation period the possibility of nephrectomy should be considered when the frequency of cyst infection reaches or exceeds two per year, or when invasive therapy or hospitalization is required. A positron emission tomography-computed tomography (PET-CT) scan can be used to determine the affected kidney and differentiate between kidney and liver cyst infections [42, 43]. In the case of yet-preserved kidney function, nephrectomy for this indication should only be considered when cyst infections lead to frequent hospitalization. In such specific cases, a mercaptoacetyl triglycine (MAG3) or dimercapto succinic acid (DMSA) scan serves as a diagnostic tool to assess separate kidney function. It sometimes occurs that patients have a disbalance in the function of both

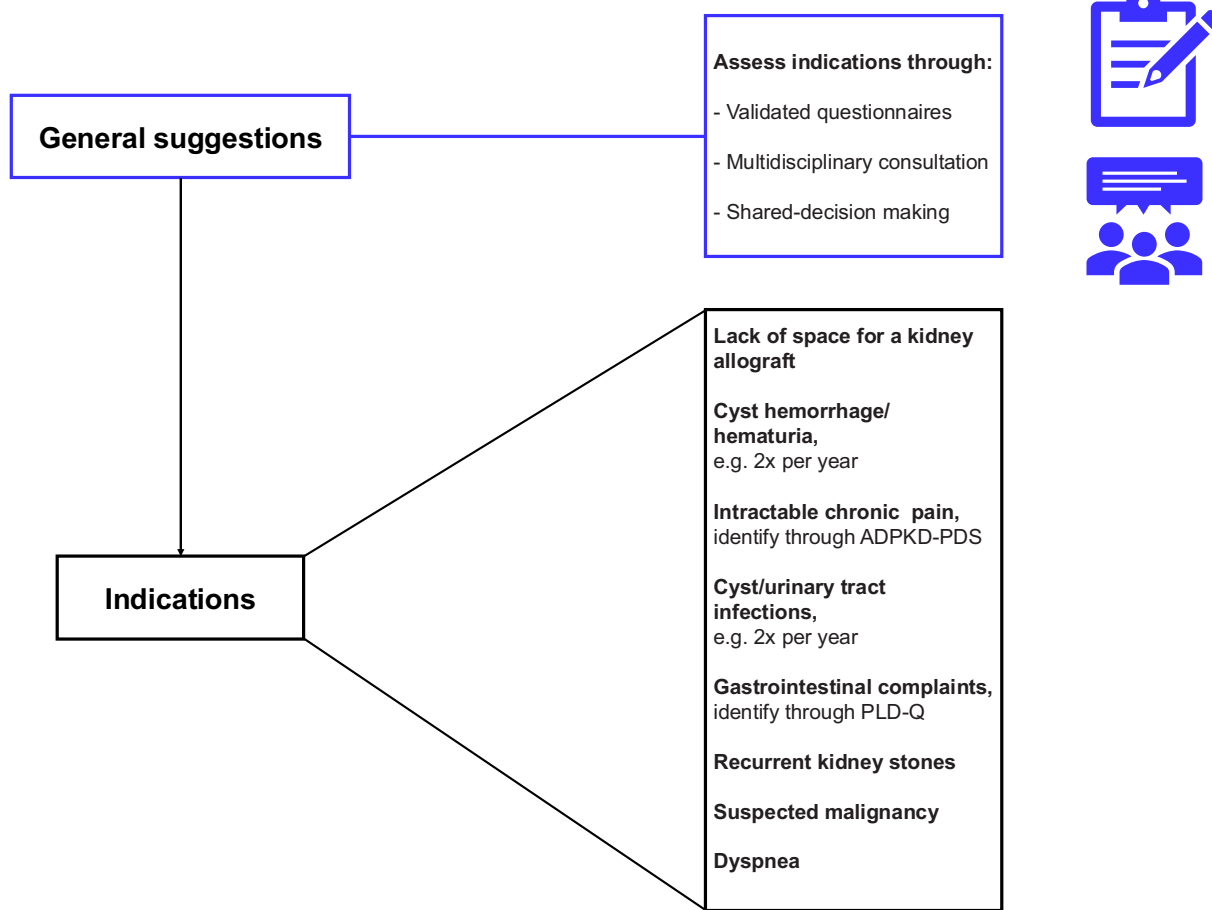


Figure 2: Visual summary of practice points concerning general suggestions and indications.

kidneys, and the dominant kidney should preferably not be removed.

A concern in transplanted patients with ADPKD is the possible increased risk of kidney cyst infections due to the use of immunosuppressive therapy. In patients with ADPKD without a kidney allograft, the incidence of cyst infections is 1 per 100 person-years [44], whereas in transplanted patients with ADPKD the incidence of cyst infection seems to be slightly increased, at 1.6 per 100 person-years [45]. The authors of the latter study found that the only risk factor for developing a cyst infection after transplantation was the history of a pre-transplantation infection [46]. Therefore, routine pre-transplantation nephrectomy to prevent cyst infections after transplantation is not typically indicated, as there appears to be no clinically relevant higher risk of cyst infection after transplantation.

Practice point 3.1: We suggest that in the workup for a kidney transplantation, a nephrectomy should be considered in case of recurrent cyst infections when the frequency of cyst infections is two or more in the last year, especially when invasive therapy or hospitalization was required to treat these cyst infections.

Practice point 3.2: Routine pre-transplantation nephrectomy to prevent cyst infections post-transplantation is not indicated in patients without a history of (recurrent) cyst infections.

When should nephrectomy be performed in cases of cyst bleedings?

Cyst hemorrhage is typically managed conservatively with hydration, pain management and, when possible, temporarily interrupting anticoagulant use [46]. When cyst hemorrhage occurs more frequently or leads to blood transfusion, nephrectomy can be considered [47–53]. To determine the affected kidney, a CT scan or cystoscopy with retrograde ureteropyelography can be performed. Considering macroscopic hematuria is also a symptom of renal malignancy [54], these steps are essential in the workup. Similar to cyst infections, there are no established guidelines regarding the exact number or severity of cyst hemorrhages requiring nephrectomy. Due to the risk of alloimmunization after blood transfusion, a severe cyst hemorrhage requiring one or multiple blood transfusions might be a more critical indication for nephrectomy than recurrent self-limiting minor cyst hemorrhages [55]. We suggest considering nephrectomy when there is frequent cyst bleeding (two or more per year), persistent cyst bleeding or a need for (multiple) blood transfusion(s). In this respect, the same argument as in cyst infections holds true: a less-functioning kidney on a MAG3 scan can lower the threshold for nephrectomy in the pre-transplantation period when patients have not yet started kidney function replacement therapy.

Practice point 4: We suggest considering nephrectomy when there is persistent cyst bleeding or recurrent cyst bleeding of two

Table 2: Timing of nephrectomy in relation to transplantation.

Publication	Nephrectomy procedures (n)	Timing	Age (years)	Sex (male), n (%)	Operative time (minutes)	Hospital stay (days)	Procedures with blood transfusions (n)	Procedures with major peri- and postoperative complications (%)	Mortality (%)
Fuller et al., 2005 [75]	7	Pre-transplantation	41.8	3 (42.9)	231 ± 14	7 ± 0.7	0	14.3	
	16	Simultaneous	37.4	9 (56.3)	370 ± 24	8.6 ± 1.2	2		
	9	Post-transplantation	49.4	4 (44.4)	208 ± 14	6.3 ± 0.6	0	11.1	11.1
Ismail et al., 2005 [51]	7	Pre-transplantation	43.7 ± 8.7	6 (85.7)	522 ± 78	8.2 ± 1.2			
	11	Simultaneous	46.3 ± 11	9 (81.8)	456 ± 66	11.7 ± 15.3		27.3	
Wagner et al., 2007 [48]	15	Pre-transplantation*	51 ± 9.5	9 (60.0)	252 ± 42	11.8 ± 2.0		33.3	
	17	Simultaneous	54 ± 6.8	8 (47.1)	430 ± 44	6.9 ± 3.2		17.6	
Kirkman et al., 2011 [4]	10	Pre-transplantation unilateral	51.5 (43–65) ^a					10.0	
	10	Pre-transplantation bilateral						60.0	20.0
	3	Sandwich technique**							
	2	Post-transplantation unilateral						50.0	
	10	Post-transplantation bilateral						40.0	10.0
Martin et al., 2012 [99]	22	Pre-transplantation*	53 (38–67)	12 (54.5)	345 (240–568)	7 (4–12)		22.7	
	15	Simultaneous	53 (43–63)	7 (46.7)	372 (278–424)	5 (3–7)		20.0	
Chebib et al., 2015 [5]	79	Pre-transplantation	50.3 ± 9.1	22 (62.9)		6 (3–14)	14	48.5 ^b	
	35	Post-transplantation	50.6 ± 9	41 (52.4)		4 (1–14)	9	26.6 ^b	
Veroux et al., 2016 [94]	25	Pre-transplantation*	44 ± 8	15 (60.0)	165 ± 44.5	15.4 ± 12.5	5	12.0	
	40	Simultaneous	51 ± 9.7	26 (65.0)	193.2 ± 71.7	13.3 ± 6.1	2	7.5	
Grodstein et al., 2017 [95]	27	Pre-transplantation*	49.9	12 (44.4)				14.7	
	161	Simultaneous	50.6	100 (62.1)				19.8	
Anselmo et al., 2019 [77]	46	Pre-transplantation	55.5 (40–70)	29 (67.5)		5 (3–15)			
	6	Post-transplantation	54.1 (40–71)	1 (16)		7.5 (4–18)			
Maxeiner et al., 2019 [27]	89	Pre-transplantation	53.92	62 (69.7)	175	7		13.5	3.4
	32	Post-transplantation	53.75	22 (68.8)	170.5	6		3.1	
Castelleijn et al., 2022 [6]	135***	Pre-transplantation	54 ± 8	71 (62.3)	155 [127–212]	10 [7–12]	10	18.3	
	37***	Post-transplantation	53 ± 7	17 (85.0)	187 [157–229]	6 [5–9]	1	8.1	
Rasmussen et al., 2022 [32]	10	Pre-transplantation*	57 ± 8.4		382 ± 56	14.5 ± 3.2	4	30.0	
	18	Simultaneous	56 ± 8.4		338 ± 59	8.1 ± 3.7	9	5.6	

Data are expressed as mean ± SD, mean [range], median [interquartile range], or median (range).

*Operative data of pre-transplantation nephrectomy were reported together with transplantation performed at a later date.

**In patients with the sandwich technique a nephrectomy was performed pre- and post-transplantation.

***Ten patients had a nephrectomy both pre and post-transplantation.

^aAge was given for all patients together.

^bMajor and minor complications were given together.

Major complications were Clavien–Dindo grade ≥ 3.

or more in the last year, especially when there was a need for blood transfusion or hospitalization.

How should kidney volume be measured when considering nephrectomy?

The main indication for pre-transplantation nephrectomy is lack of space for the future kidney allograft. To assess whether the size of the polycystic kidneys and the enlarged cystic liver might pose problems, the volume of the kidneys and liver should be determined. Volume-related complaints should be assessed by medical history, physical examination and/or imaging, such as ultrasound, CT and MRI [56]. Physicians indicated that their preferred methods for determining the lack of space for a kidney allograft are abdominal MRI or CT (82.1%), and physical examination (58.9%). It is important to note that the level of complaints caused by a high combined kidney and liver volume depends on body length and body type. For instance, large kidneys are more of a problem in small than in tall people. Moreover, patients with well-developed abdominal muscles will have more complaints from a high combined total kidney and liver volume due to less space to expand. To assess the kidney's relation to the iliac crest, physical examination can be used. For evaluation of kidney volume, MRI is usually preferred due to its high sensitivity and specificity, whereas ultrasound assessment is physician-dependent and less reliable for kidney volume measurement. CT imaging has the disadvantage of radiation and requires contrast infusion for adequate assessment of the vessels, which is relatively contraindicated because of the possible impact on kidney function in later-stage CKD. On the other hand, it is the preferred technique for evaluation of calcification and patency of the iliac arteries to assess surgical options for a kidney transplantation. If recent CT imaging is already available, there is no need for additional evaluation by MRI.

To our knowledge, there is no volume-related cut-off value to decide when nephrectomy is needed. However, if the native kidneys extend below the iliac crest, which is an indication that the intra-abdominal space is limited, it may be reasonable to consider nephrectomy to create space for a kidney allograft. This indication can be further investigated through clinical examination. Depending on the distribution of the kidney cysts (e.g. single large cyst) and the condition of the patient, other volume-reducing therapies (e.g. cyst fenestration or embolization) can also be considered. Renal artery embolization will be discussed in more detail in a later paragraph. (Serial) measurement of total kidney and liver volume is an important step in identifying patients who may benefit from nephrectomy. Patients with ADPKD and a high combined kidney and liver volume develop symptoms only gradually; they may not appreciate the seriousness of their condition due to habituation.

Several studies observed that kidney volume decreases when patients reach kidney failure and become kidney replacement therapy-dependent. During follow-up up to 10 years post-transplantation, kidney volume reduction ranged from 20% to 46% [57, 58]. Similar results were reported in patients on dialysis [59]. However, this is not the case for all patients; a limited number of patients can still experience an increase in kidney volume. Given that kidney volume generally decreases after transplantation, routine pre-transplantation nephrectomy to prevent new volume-related complaints after transplantation is not advised. If a patient continues to experience volume-related complaints post-transplantation, an MRI or CT can be performed to assess kidney growth as well as liver volume and growth. When there is persistent kidney growth, a nephrectomy can effectively reduce volume-related

complaints and can, therefore, be considered. Validated quality of life questionnaires may help to identify and objectify complaints. The questionnaires will be discussed in the next section.

Practice point 5.1: If there is no recent adequate imaging available, we suggest performing an MRI or CT when considering a nephrectomy during the pre-transplantation evaluation to determine the size of the kidneys and liver.

Practice point 5.2: If the native kidneys extend to below the iliac crest, a nephrectomy or another form of volume-reducing therapy should be considered to create space for a kidney allograft, with additional input from clinical examination.

Practice point 5.3: Given that, on average, there is a decrease in volume of the polycystic kidney after onset of kidney failure, routine pre-transplantation nephrectomy to prevent new volume-related gastrointestinal complaints post-transplantation is not advised.

Practice point 5.4: To identify patients with post-transplantation kidney growth, we suggest performing an abdominal MRI or CT in patients who continue to have or develop potentially volume-related complaints during this period.

Practice point 5.5: We suggest performing a nephrectomy in patients with persisting post-transplantation kidney growth and severe volume-related complaints.

How are complaints assessed in relation to nephrectomy?

Patients with ADPKD often experience symptoms such as abdominal fullness, nausea, loss of appetite, and pain [5]. Chronic pain is a common symptom in ADPKD, with a prevalence up to 50% [60]. To comprehensively evaluate pain complaints and determine their relation to ADPKD, a detailed medical history, physical examination, and kidney imaging are recommended [61]. Pain can further be assessed using a visual analog scale or a validated questionnaire. Suitable questionnaires include the Short Form-36, Short-Form McGill Pain Questionnaire, Multidimensional Pain Inventory, ADPKD-Pain and Discomfort Scale (PDS), or ADPKD-Impact Scale (IS) [62–66]. The Short Form-36 is a validated questionnaire for assessment of health-related quality of life, while the Short-Form McGill Pain Questionnaire and Multidimensional Pain Inventory are specifically for pain assessment. Both the ADPKD-IS and ADPKD-PDS are validated questionnaires in patients with ADPKD. The ADPKD-PDS was created to assess pain and fullness complaints and contains 20 items in which patients can score complaints from 1 (mild) to 5 (severe), resulting in a total severity score between 1 and 5. An ADPKD-PDS score of ≥ 3 represents the 16% of patients with the most severe pain complaints [67]. If an overall pain and discomfort score of ≥ 3 on the ADPKD-PDS is scored, a nephrectomy can be considered. In clinical practice, physicians assess pain complaints for potential nephrectomy indications using general questions (51.8%), spontaneously reported complaints (39.3%), physical examination (37.5%), or a review of systems (32.1%).

Besides pain, gastrointestinal symptoms are common in ADPKD, affecting ~61.2% of patients [68]. Validated questionnaires such as the GI (gastrointestinal) symptom questionnaire or the Polycystic Liver Disease Questionnaire (PLD-Q) (primarily used for PLD complaints) can be used to assess gastrointestinal complaints and results in a score between 0 and 100, with a higher score representing more complaints

[69, 70]. A score of ≥ 33 on the PLD-Q represents the 25% of patients with the most severe gastrointestinal symptoms and in these patients a nephrectomy can therefore be considered [70]. Based on findings from the questionnaire, physicians typically assessed gastrointestinal complaints using a general question (46.4%) or based on spontaneously reported complaints (42.9%).

Disturbed self-image or embarrassment, as a result of ADPKD-related increase in abdominal volume, can be observed, especially in women [71, 72]. In women, liver volume increases relatively more due to estrogen stimulation [73]. The frequency of physicians' inquiries about this issue is not well documented in the literature. Eighty-nine percent of physicians in the questionnaire indicated that they did not ask about this, suggesting that a disturbed self-image is an unrecognized problem. Paying attention to the emotional and psychological impact of ADPKD is essential. In selected cases, performing nephrectomy can improve quality of life.

To summarize, while many complaints are assessed using general questions or spontaneously reported complaints, it is likely that slowly developing complaints may go unnoticed with this approach. Therefore, we suggest using the ADPKDPDS to determine whether there are any pain and discomfort complaints, and PLD-Q to determine whether there are any gastrointestinal and fullness complaints in all patients with ADPKD in the pre-transplantation period. This method can objectively assess whether there may be an indication for nephrectomy that would have been missed when the only assessment used was a general question concerning complaints.

Practice point 6: We suggest using validated questionnaires to objectively assess whether potential indications for nephrectomy exist, such as the ADPKD-PDS to assess pain and discomfort, and the PLD-Q to assess gastrointestinal and fullness complaints.

What is the best timing for nephrectomy?

In the literature the timing of nephrectomy is mainly described in relation to kidney transplantation, with options being pre-transplantation, combined with transplantation, or post-transplantation [74–77]. Table 2 provides an overview of the studies used in the meta-analysis of timing. Analysis of pre-transplantation versus simultaneous nephrectomy indicates that simultaneous nephrectomy is more common when a living donor allograft is available (RD -0.45 [95% CI -0.88 to -0.01]) (Supplementary Fig. S1).

During a combined procedure, the unilateral native kidney is removed before graft implantation. This could increase the surgery time by 1–3 hours, with consequences for the cold ischemia time when the surgeries are not performed in parallel. This can become problematic in cases of postmortem kidney donation with an already long-lasting procedure. In some cases, hemodynamic instability might occur during nephrectomy, and consequently cancellation of the transplantation could happen. In the case of living donation, simultaneous surgeries can be organized more easily. While simultaneous nephrectomy and transplantation with a deceased donor has been performed [8], the logistical challenges, especially during the evenings and nights, make it less feasible for most hospitals. Of note, no differences were found between pre-transplant and simultaneous procedures in operative time (MD -46.7 [95% CI -149.3 to 56.0]), hospital stay (MD 2.3 [95% CI -1.4 to 5.9]), need for blood transfusions (RD 0.01 [95% CI -0.40 to 0.42]), and incidence of major

complications (Clavien–Dindo class ≥ 3) (RD 0.03 [95% CI -0.10 to 0.16]) (Figs 3–5, Supplementary Fig. S2).

When comparing nephrectomy procedures performed pre-transplantation or post-transplantation, a lower risk of major complications was observed in post-transplantation procedures (RD 0.11 [95% CI 0.04 – 0.18]). This could result from an improved patient physical condition after transplantation. No differences were found in operative time (MD 4.7 [95% CI -41.6 to 51.0]), hospital stay (MD 1.4 [95% CI -0.5 to 3.3]), incidence of procedures with blood transfusions (RD 0.12 [95% CI -0.28 to 0.52]), and living versus deceased donors (RD -0.03 [95% CI -0.35 to 0.29]) (Figs 3–5, Supplementary Fig. S2).

In addition, there is no difference in patient and graft survival between pre-transplantation and post-transplantation nephrectomy [5, 6]. Possible outliers and analyses with an $I^2 > 50\%$ were assessed as a sensitivity analysis. There were no major differences found.

Another point of debate regarding the timing of pre-transplantation nephrectomy is the interval between a nephrectomy for the indication lack of space for a kidney allograft and the transplantation. In the questionnaire, there was no consensus; $\sim 44\%$ preferred performing a nephrectomy just before kidney replacement therapy (when a central venous catheter for dialysis can be placed during anesthesia), while 39.3% preferred doing it after the start of kidney replacement therapy. In the case of a staged procedure before transplantation, the majority of physicians indicated that, in general, they deemed a patient ready for transplantation after 1–2 months (55.4%) or alternatively 2–4 months (32.1%). It should be noted that conducting nephrectomy before kidney failure has been reached will significantly reduce kidney function, expedite the onset of kidney failure and therefore impact morbidity and mortality. Performing the procedure when patients are not close to reaching kidney failure should only be considered in situations where no alternative treatment options exist. In addition, performing nephrectomy in patients on dialysis reduces the residual urine diuresis, which is related to a decrease in quality of life [78]. Therefore, this decision should be taken with caution.

The timing of nephrectomy should be determined based on the specific indication and the availability of a living donor. When there is lack of space for a kidney allograft, we suggest performing nephrectomy just before (1–3 months) or simultaneously with transplantation, with simultaneous nephrectomy being more feasible in well-selected patients with a living donor. The findings from the meta-analysis also indicate that post-transplantation nephrectomy carries no higher risk of complications than pre-transplantation nephrectomy. Therefore, nephrectomy before transplantation is not recommended unless there is a clear indication. A visual summary of practice points concerning timing is given in Fig. 6.

Practice point 7.1: The timing of nephrectomy in relation to a kidney transplantation should be determined based on the specific indication and the availability of a living donor allograft.

Practice point 7.2: Performing a nephrectomy before reaching kidney failure should be reserved for cases where there are no other treatment options available. Nephrectomy during this phase of the disease will reduce kidney function, expediting the onset of kidney failure and potentially negatively impacting morbidity and mortality.

Practice point 7.3: When lack of space for a kidney allograft is the indication for nephrectomy as determined by the transplant

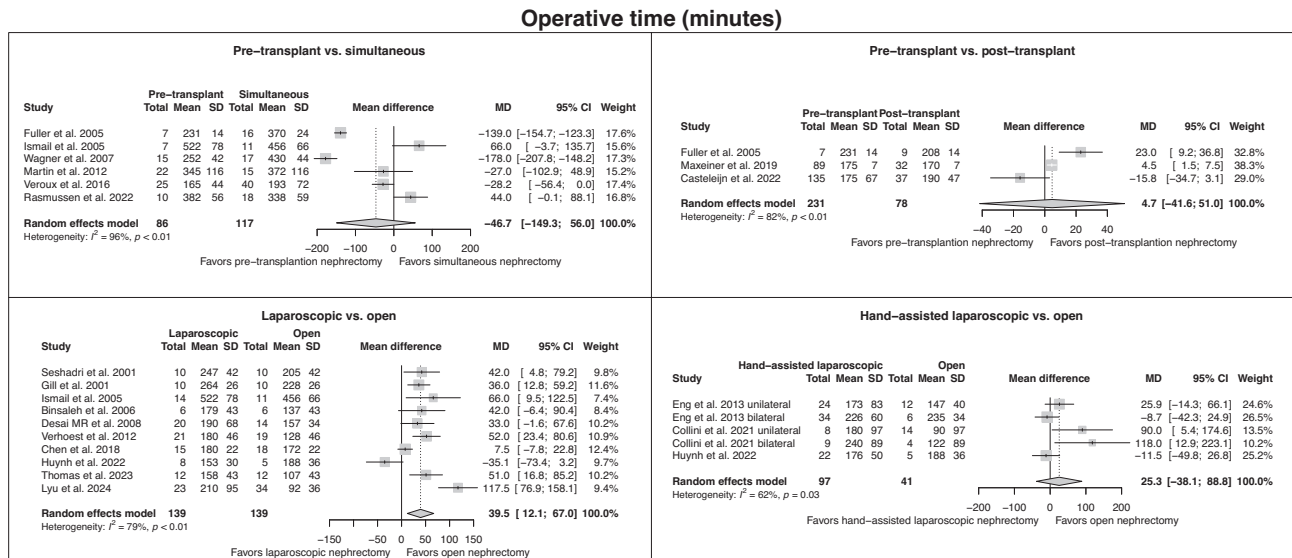


Figure 3: Meta-analysis of studies investigating operative time in minutes of nephrectomies performed pre-transplantation versus during transplantation (left upper panel), pre-transplantation versus post-transplantation (right upper panel), when the nephrectomy is performed via a laparoscopic procedure versus an open procedure (left bottom panel) or via a hand-assisted laparoscopic procedure versus an open procedure (right bottom panel).

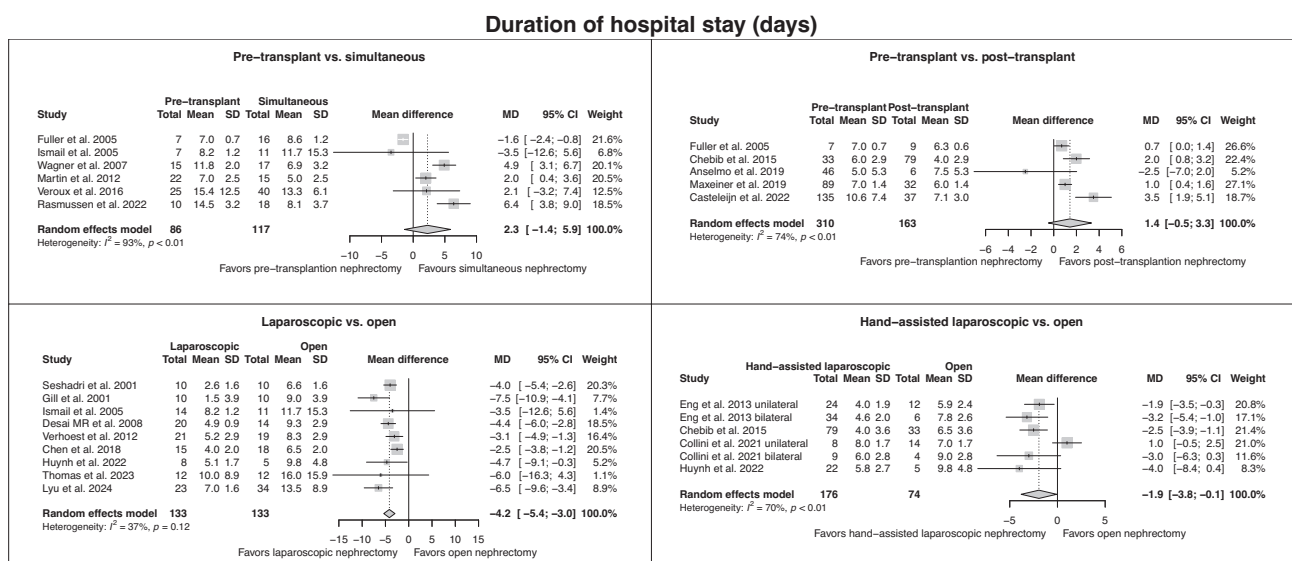


Figure 4: Meta-analysis of studies investigating the duration of hospital stay in days of nephrectomies performed pre-transplantation versus during transplantation (left upper panel), pre-transplantation versus post-transplantation (right upper panel), when the nephrectomy is performed via a laparoscopic procedure versus an open procedure (left bottom panel) or via a hand-assisted laparoscopic procedure versus an open procedure (right bottom panel).

surgeon, the preferred timing for nephrectomy is just before (1–3 months) or simultaneously with transplantation, to minimize the time with reduced residual kidney function, with simultaneous nephrectomy being more feasible in well-selected patients with a living donor in centers with experience with this procedure.

Which surgical approach may be the preferred choice?

Several surgical approaches can be used to perform nephrectomy in patients with ADPKD, such as open, and minimally invasive

hand-assisted laparoscopic, pure laparoscopic or robot-assisted laparoscopic surgery [79–82]. The questionnaire showed that physicians commonly use an open (40.5%) or a transperitoneal laparoscopic (21.6%) approach. Laparoscopic approaches become more challenging with higher kidney volumes [83]. For this reason, several papers discussed a hand-assisted laparoscopic approach, which involves the use of a hand-port to assist in the procedure [33, 84, 85]. Several retrospective studies have compared open versus laparoscopic procedures and open versus hand-assisted laparoscopic procedures, and these studies were included in the meta-analysis (Table 3).

Table 3: Comparison of surgical techniques that are used for nephrectomy of a native polycystic kidney.

Publication	Nephrectomy procedures (n)	Surgical technique	Age of patients (years)	Sex (male), n (%)	Operative time (minutes)	Hospital stay (days)	Procedures with blood transfusions (n)	Procedures with major peri- and postoperative complications (%)	Mortality (%)
Seshadri et al., 2001 [87]	10	Laparoscopic	47 [37–60]	8 (80.0)	247 (150–420)	2.6 (1–4)	0	10.0	
	10	Open	51 [40–63]	7 (70.0)	205 (100–270)	6.6 (5–11)	1		
Gill et al., 2001 [7]	10	Laparoscopic	53 [39–63]	6 (60.0)	264 (204–426)	1.5 (1–7)	0 (0–6)	30.0	
	10	Open	49 [37–56]	8 (80.0)	228 (180–330)	9 (7–13)	1 (0–4)	20.0	
Ismail et al., 2005 [51]	14	(Hand-assisted)* laparoscopic	43.7 ± 8.7	6 (85.7)	522 ± 78	8.2 ± 1.2			
	11	Open	46.3 ± 11	9 (81.8)	456 ± 66	11.7 ± 15.3		27.3	
Binsaleh et al., 2006 [53]	6	Laparoscopic	52 [42–67]	4 (66.7)	179 [113–240]	4.7 [2–11]	0	16.7	
	6	Open	52 [43–64]	4 (66.7)	137 [90–190]	5 [3–8]	0		
Desai MR et al., 2008 [76]	20	Laparoscopic	49 ± 6	11 (84.6)	190 ± 67.6	4.86 ± 0.9	0.9 ± 0.6	35.0	
	14	Open			157 ± 34	9.26 ± 2.9	1.3 ± 0.5	42.9	
Verhoest et al., 2012 [15]	21	Laparoscopic	53 [41–71]	8 (38.1)	180 [90–310]	5.2 [3–11]	1	9.5	
	19	Open	53 [40–71]	10 (52.6)	128 [100–170]	8.28 [5–24]	3	36.8	
Eng et al., 2013 [13]	24	Hand-assisted laparoscopic, unilateral	51 ± 8**	33 (56.9)	173.0 ± 82.8	4.0 ± 1.9	4	16.7	
	12	Open, unilateral	49 ± 8**	13 (72.2)	147.1 ± 40.2	5.9 ± 2.4	5	8.3	
	34	Hand-assisted laparoscopic, bilateral			226.0 ± 59.5	4.6 ± 2.0	9	32.4	
Chebib et al., 2015 [5]	6	Open, bilateral			234.7 ± 33.7	7.8 ± 2.6	5	33.3	
	79	(Hand-assisted) laparoscopic				4 [1–14]	10	33.0 ^a	
	33	Open				6.5 [3–14]	13	33.3 ^a	
Chen et al., 2018 [100]	15	Laparoscopic	49.0 [42.0–61.0]	8 (53.3)	180.0 [153.0–287.0]	4.0 [3.0–6.0]	1	6.7	
	18	Open	51.5 [47.8–56.0]	12 (66.7)	172.5 [122.8–249.8]	6.5 [5.0–10.3]	6	11.1	
Collini et al., 2021 [33]	8	Hand-assisted laparoscopic, unilateral	54 (35–63)	4 (50.0)	180 (105–270)	8 (4–12)	1	12.5	
	14	Open, unilateral	55 (38–67)	6 (42.8)	90 (60–135)	7 (4–13)	1	7.1	
	9	Hand-assisted laparoscopic, bilateral	54 (39–61)	5 (55.5)	240 (125–285)	6 (5–7)	1	22.2	
Huynh et al., 2022 [79]	4	Open, bilateral	52 (50–54)	3 (75.0)	122 (105–150)	9 (5–9)	2		
	22	Hand-assisted laparoscopic	48.9 ± 7.8	16 (72.7)	176.5 ± 50.2	5.8 ± 2.7	1	9.1	
	8	Laparoscopic	50.5 ± 9.8	3 (37.5)	152.9 ± 30.3	5.1 ± 1.7	0		
	5	Open	53.0 ± 8.3	5 (100)	188.0 ± 36.5	9.8 ± 4.8	2	20.0	
Thomas et al., 2023 [86]	12	Laparoscopic	60 [39–68]	6 (66.7)	158 [85–227]	10 [6–35]		8.3	
	12	Open	55 [34–68]	9 (75.0)	107 [56–174]	16 [4–56]		8.3	
Lyu et al., 2024 [81]		Laparoscopic	51.8 ± 9.5	14 (60.9)	210 [130–250]	7 [6–8]		4.3	
		Open	47.5 ± 6.9	22 (64.7)	93 [84–130]	14 [9–21]		2.9	

Data are expressed as mean ± SD, mean [range], median [interquartile range], or median (range).

*Laparoscopic nephrectomy was performed hand-assisted in 3 patients and non-hand-assisted in 4 patients.

**Age and gender were given for uni- and bilateral nephrectomy together.

^aMajor and minor complications were given together.

Major complications were Clavien–Dindo grade ≥3.

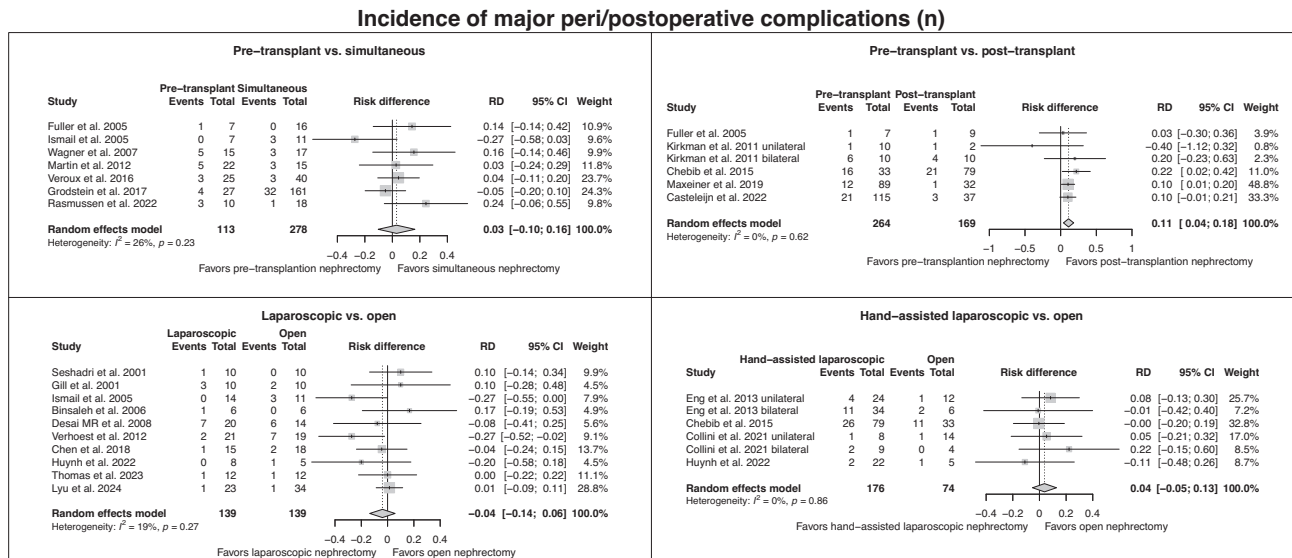


Figure 5: Meta-analysis of studies investigating the incidence of major peri-/postoperative complications of nephrectomies performed pre-transplantation versus during transplantation (left upper panel), pre-transplantation versus post-transplantation (right upper panel), when the nephrectomy is performed via a laparoscopic procedure versus an open procedure (left bottom panel) or via a hand-assisted laparoscopic procedure versus an open procedure (right bottom panel).

When comparing open versus laparoscopic procedures, it was found that laparoscopic nephrectomy resulted in a significantly longer operative time (MD 39.5 min [95% CI 12.1–67.0]) but a shorter hospital stay (MD –4.2 days [95% CI –5.4 to –3.0]) (Figs 3 and 4). Importantly, there was no difference in the incidence of patients requiring blood transfusions, the occurrence of major complications, or the weight of the native kidney after resection (Fig. 5, Supplementary Figs S1 and S3). One publication from 2023 reported that in 24 procedures the quality of life tended to be better after laparoscopic nephrectomy compared with open nephrectomy [86].

When comparing open versus hand-assisted laparoscopic nephrectomy, hand-assisted laparoscopic procedures were associated with a significantly shorter hospital stay (MD –1.9 days [95% CI –3.8 to –0.1]) and a lower incidence of patients requiring blood transfusions (RD –0.27 [95% CI –0.49 to –0.04]) (Fig. 4, Supplementary Fig. S2). However, there were no significant differences in operative time and the incidence of major complications during procedures (Figs 3 and 5). Sensitivity analyses showed similar results. Of note, hand-assisted laparoscopic procedures were more commonly performed in patients with a lower kidney weight after resection (MD –1377 mL [95% CI –1790 to –964]) (Supplementary Fig. S4). The variation in kidney weight after resection in hand-assisted laparoscopic procedures can be attributed to the aspiration or rupture of cysts to reduce the kidney volume and weight after resection [87]. It is important to note that aspiration or rupture of cysts is not advised in cases of nephrectomy for malignancy in order to prevent local tumor spread [13]. Kidney volume may play a role in the choice of open versus laparoscopic procedure. Lipke et al. reported a higher risk for conversion in patients with a kidney volume exceeding 3500 mL [88]. In contrast, Abrol and Prieto found no need for conversion in 51 simultaneous bilateral hand-assisted laparoscopic procedures with a median total kidney volume of 5100 mL [89]. These data indicate that it is possible to perform a hand-assisted laparoscopic nephrectomy even in patients with large polycystic kidneys [89]. Nevertheless, it is important to note that even with a hand-

assisted procedure a larger incision is needed for the extraction of the native kidney, especially in the case of larger kidney volumes.

The laparoscopic technique can also be performed using a robot-assisted device [80, 90–92]. Four case series have described a total of 26 robot-assisted procedures, five of them being performed concurrently with a kidney transplantation. Overall, the peri- and postoperative outcomes in these publications were comparable to standard laparoscopic procedures. It is important to note that, as with laparoscopic procedures, robot-assisted procedures are more difficult in patients with larger kidneys. Furthermore, robot-assisted procedures result in higher hospital costs compared with regular laparoscopic procedures [93].

The choice of surgical approach for nephrectomy is dependent on the surgeon's experience and the availability of (robot-assisted) laparoscopy. According to the meta-analysis, both the open and laparoscopic approaches demonstrate relative safety. However, when a surgeon has sufficient experience with (robot-assisted) laparoscopic nephrectomy, this may be the preferred approach, especially in cases involving relatively smaller polycystic kidneys because the (robot-assisted) laparoscopic technique yielded several advantages. These advantages included a significantly shorter hospital stay, a comparable rate of major complications, and a lower incidence of blood transfusions. It is important to emphasize the desirability of avoiding blood transfusions, as they carry a risk of alloimmunization [55]. A visual summary of practice points concerning surgical approach, lateralization, outcomes, and renal embolization is given in Fig. 7.

Practice point 8.1: The choice of a specific surgical approach for nephrectomy (laparotomy versus laparoscopy, robot-assisted or not) is dependent on kidney size, the surgeon's experience and the availability of facilities for performing (robot-assisted) laparoscopy.

Practice point 8.2: A laparoscopic nephrectomy is generally preferred over an open approach, while taking into account the aforementioned considerations.

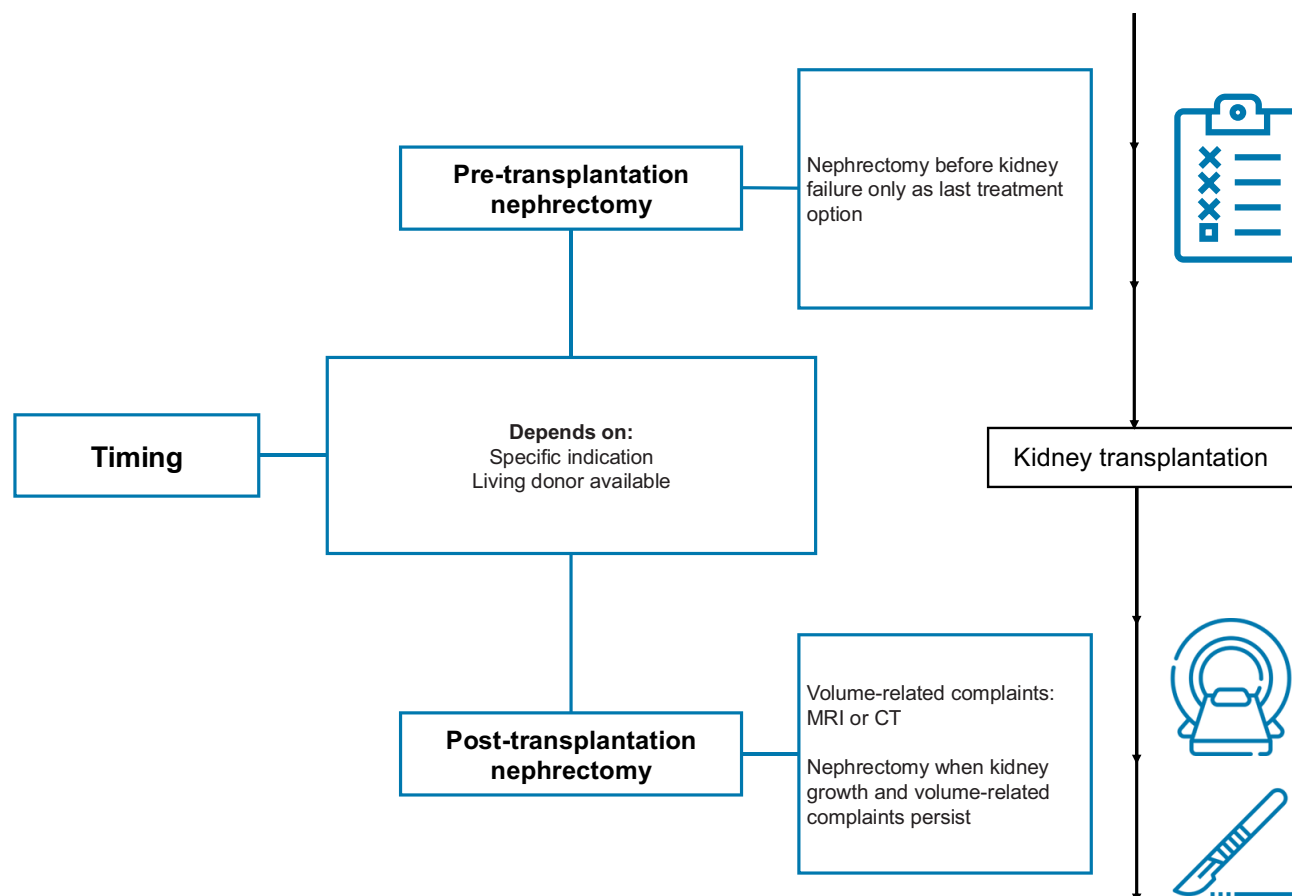


Figure 6: Visual summary of practice points concerning timing.

What are the complications of nephrectomy?

For the assessment of complications and mortality, a total of 2128 procedures were analyzed. Peri- and postoperative complications were noted in 35.1% of cases (Table 4). The most common minor complications were wound infection, need for blood transfusion, fever, pneumonia, and prolonged ileus [5, 47, 49, 94–97]. The incidence of major complications was 14.7% and consisted of wound dehiscence, arteriovenous fistula thrombosis, incisional hernias, intra-abdominal hemorrhage, sepsis, intra-abdominal organ injury, myocardial infarction, and lymphocele [7, 10, 13, 48, 98].

In most publications, there was no or low postoperative mortality [4, 5, 13, 15, 27, 33, 48, 75, 95, 99]. Analysis of 2106 procedures found a mortality rate of 1.5% (Table 4). The mortality rate in patients with ADPKD is primarily driven by postoperative death after emergency procedures [38]. Studies comparing survival of patients who underwent both nephrectomy as well as transplantation versus those who underwent transplantation alone consistently reported similar overall survival rates between the two groups [5, 6, 23, 99]. In addition, similar graft survival rates were observed in these studies. In cases of an emergency procedure, as expected, the incidence of complications is higher [26, 38]. Data from the questionnaire indicated that 83.6% of the participants regarded a nephrectomy as an intermediate-risk procedure, while 67.9% considered it moderately burdensome for patients (Supplementary Table S4).

To summarize, these findings suggest that nephrectomy represents an intermediate risk procedure with an acceptable

but not inappreciable mortality rate. Patient and graft survival rates of combined or separate nephrectomy and transplantation have been demonstrated to be equivalent with transplantation alone.

Statement 9.1: Nephrectomy represents an intermediate risk procedure with an acceptable but not inappreciable mortality rate.

Statement 9.2: In selected cases, patient and graft survival rates of combined or separate nephrectomy and transplantation have been described to be on a par with transplantation alone.

Unilateral or bilateral nephrectomy?

In cases in which nephrectomy is deemed necessary due to space constraints or to alleviate gastrointestinal complaints related to a high intra-abdominal volume, a clinical dilemma arises concerning whether one or both kidneys should be removed. It is commonly assumed that a bilateral procedure has more risks compared with a unilateral one. This assumption is supported by a study by Lucas *et al.*, who found a higher risk of blood loss and complications in a bilateral procedure [9]. Similar findings were observed by Kirkman *et al.* and Chen *et al.*, who reported a higher rate of postoperative complications and mortality [4, 100]. In addition, there are several case reports describing a decompensated liver failure in patients with ADPKD and a massive polycystic liver after bilateral nephrectomy [101, 102]. Bilateral nephrectomy might also exacerbate hypotension through loss of the intrarenal renin-angiotensin system after

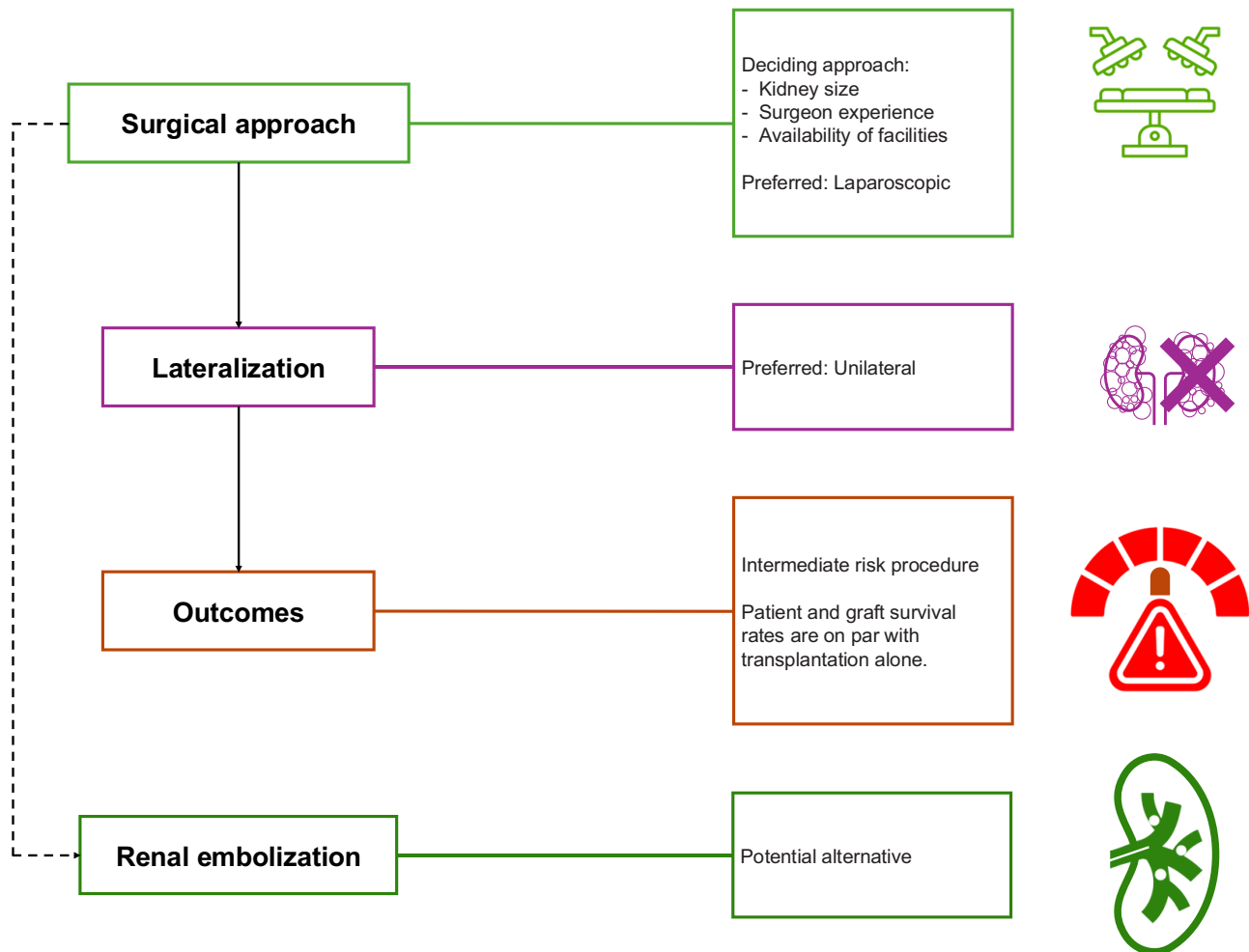


Figure 7: Visual summary of practice points concerning surgical approach, lateralization, outcomes and renal embolization.

the procedure. Consequently, advanced hemodynamic monitoring may be indicated. On the other hand, data published in 2023 showed a better physical quality of life after bilateral versus unilateral nephrectomy [36]. This suggests that patients who experience severe physical complaints may benefit from bilateral nephrectomy. Results from the questionnaire indicated that most physicians (91.1%) preferred to perform a unilateral nephrectomy. Considering the higher risk of complications in bilateral nephrectomy, we suggest that a unilateral nephrectomy is the preferred choice unless there is a clear reason to perform it bilaterally.

The next question then arises: Which kidney should be removed? When there is insufficient space for transplantation, the prevailing suggestion is to perform a right-sided nephrectomy due to the more superficial position of the blood vessels in the right iliac fossa for graft anastomosis [4, 22, 94]. Nonetheless, if the left kidney is significantly larger than the right kidney, a left-sided nephrectomy is preferred for this indication. The effect of lateralization of nephrectomy on alleviation of gastrointestinal complaints has never been investigated. We hypothesize that when there is abdominal fullness, nausea, and the inability to eat large portions, a left-sided nephrectomy may be preferred to relieve abdominal pressure on the stomach, especially in patients who also have (left-sided) enlarged polycystic livers. Fifty percent of physicians surveyed had no preference for either the left or

right kidney, while one-third (33.9%) favored the right kidney, and the remaining 16.1% favored the left kidney.

In conclusion, in cases of spatial constraints for transplantation, we suggest performing a unilateral right-sided nephrectomy. When there are additional complaints in these patients that could be alleviated by a left-sided nephrectomy, for instance gastrointestinal, we suggest performing a nephrectomy on the affected side.

Practice point 10.1: Considering the higher risk of complications when performing bilateral nephrectomy, we suggest that a unilateral nephrectomy is the preferred choice, unless there are specific indications to perform a bilateral nephrectomy.

Practice point 10.2: In cases of spatial constraints for transplantation, we suggest performing a unilateral right-sided nephrectomy unless there are specific complaints or other reasons that necessitate a left-sided (or bilateral) nephrectomy.

Is renal embolization an alternative to nephrectomy?

Renal artery embolization (RAE) has emerged as an alternative to nephrectomy for symptom relief and volume reduction. This procedure is based on the induction of an occlusion of the renal artery, which leads to infarction, and subsequent renal atrophy,

Table 4: Peri-/postoperative complications and mortality.

Publication	Number of procedures	Procedures with any peri-/postoperative complication, n (%)	Procedures with major peri-/postoperative complication, n (%)	Mortality, n (%)
Bennett et al., 1973 [47]	31	8 (25.8)	4 (12.9)	1 (3.2)
Glassman et al., 2000 [74]	10	2 (20.0)	2 (20.0)	0 (0.0)
Seshadri et al., 2001 [87]	20	4 (20.0)	1 (5.0)	0 (0.0)
Gill et al., 2001 [7]	20	9 (45.0)	3 (15.0)	0 (0.0)
Bendavid et al., 2004 [52]	22	11 (50.0)	6 (27.3)	0 (0.0)
Fuller et al., 2005 [75]	32	2 (6.3)	2 (6.3)	1 (3.1)
Rozanski et al., 2005 [96]	30	6 (20.0)	5 (16.7)	0 (0.0)
Ismail et al., 2005 [51]	16	7 (43.8)	3 (18.8)	0 (0.0)
Whitten et al., 2006 [84]	10	2 (20.0)	2 (20.0)	0 (0.0)
Binsaleh et al., 2006 [53]	12	0 (0.0)	0 (0.0)	0 (0.0)
Lipke et al., 2007 [88]	18	5 (27.8)	1 (5.6)	0 (0.0)
Nunes et al., 2007 [23]	16	1 (6.3)	1 (6.3)	0 (0.0)
Wagner et al., 2007 [48]	32	9 (28.1)	2 (6.3)	0 (0.0)
Binsaleh et al., 2008 [12]	16	3 (18.8)	3 (18.8)	0 (0.0)
Desai MR et al., 2008 [76]	34	14 (41.2)	13 (38.2)	0 (0.0)
Desai PJ et al., 2008 [11]	12	3 (25.0)	2 (16.7)	0 (0.0)
Kramer et al., 2009 [25]	20	4 (20.0)	4 (20.0)	0 (0.0)
Lucas et al., 2010 [9]	42	10 (23.8)	4 (9.5)	0 (0.0)
Kirkman et al., 2011 [4]	35	12 (34.3)	12 (34.3)	3 (8.6)
Patel et al., 2011 [26]	31	20 (64.5)		1 (3.2)
Skauby et al., 2012 [49]	78	34 (43.6)	26 (33.3)	1 (1.3)
Verhoest et al., 2012 [15]	40	20 (50.0)	9 (22.5)	0 (0.0)
Martin et al., 2012 [99]	37	17 (45.9)	8 (21.6)	0 (0.0)
Eng et al., 2013 [13]	76	20 (26.3)	18 (23.7)	0 (0.0)
Bansal et al., 2014 [98]	39	4 (10.3)	3 (7.7)	0 (0.0)
Asimakopoulos et al., 2015 [24]	19	4 (21.1)	1 (5.3)	0 (0.0)
Chebib et al., 2015 [5]	112	37 (33.0)		0 (0.0)
García-Rubio et al., 2015 [82]	27	7 (25.9)	2 (7.4)	0 (0.0)
Wisenbaugh et al., 2015 [85]	68	24 (35.3)	9 (13.2)	0 (0.0)
Kim et al., 2016 [40]	22	13 (58.5)		
Veroux et al., 2016 [94]	65	8 (12.3)	6 (9.2)	0 (0.0)
Ahmad et al., 2016 [10]	72	37 (51.4)	11 (15.3)	0 (0.0)
Benoit et al., 2016 [97]	82	24 (29.3)	10 (12.2)	0 (0.0)
Grodstein et al., 2017 [95]	188	94 (50.0)	36 (19.1)	0 (0.0)
Chen et al., 2018 [100]	33	9 (27.3)	3 (9.1)	0 (0.0)
Bellini et al., 2019 [14]	33	5 (15.2)	2 (6.1)	1 (3.0)
Maxeiner et al., 2019 [27]	121	51 (42.1)	8 (6.6)	3 (2.5)
Anselmo et al., 2019 [77]	53	16 (30.2)	3 (5.7)	2 (3.8)
Abrol and Prieto, 2020 [89]	51	16 (31.4)	4 (7.8)	0 (0.0)
Lubennikov et al., 2021 [38]	108		21 (19.4)	18 (16.7)
Collini et al., 2021 [33]	35	9 (25.7)	3 (8.6)	1 (2.9)
Casteleijn et al., 2022 [6]	152	54 (35.5)	24 (15.8)	0 (0.0)
Rasmussen et al., 2022 [32]	28	21 (75.0)	4 (14.3)	0 (0.0)
Huynh et al., 2022 [79]	35	16 (45.7)	3 (8.6)	0 (0.0)
Thomas et al., 2023 [86]	24		2 (8.3)	0 (0.0)
Masterson et al., 2023 [92]	14	2 (14.3)	0 (0.0)	0 (0.0)
Lyu et al., 2024 [81]	57	26 (45.6)	2 (14.7)	0 (0.0)
Total	2128	35.1%*	14.7%*	1.5%*

*Based on 1996, 1963, and 2106 procedures, respectively.

and gain of space [103]. Different techniques using coils, plugs, microparticles, ethanol, glue or a combination of these modalities have been described for embolization of a polycystic kidney [104, 105]. A summary of recent publications on RAE is provided in Table 5. RAE was able to reduce kidney volume by 35%–50% in 3–6 months, sufficiently reducing volume-related complaints and creating enough space for a kidney allograft in 65 (89.0%) of 73 patients with ADPKD described in a study from 2015 [104]. However, in 3.4%–28.6% of reported patients, sufficient volume reduction was not achieved due to accessory arteries or recanalization

of the renal artery [104, 106–108]. To prevent recanalization, it has been recommended to embolize with a liquid agent (i.e. glue, ethanol) or microparticles to perform a definitive distal embolization and necrosis [109]. Another study in 188 patients with a median total kidney volume of 4497 mL demonstrated a favorable impact of RAE on patients' quality of life, resulting in improvements in abdominal fullness, appetite, and nausea [110]. In patients undergoing peritoneal dialysis, treatment with RAE has been described to be associated with superior survival of the peritoneal dialysis technique, with a reduced requirement

Table 5: Recent publications on renal artery embolization in patients with ADPKD.

Publication	Patients (n)	Kidneys (n)	Indication of RAE	Technical failure, n (%)	Duration of hospital stay, days	Post-embolization syndrome, n (%)	Major complications, n (%)	1-year mortality rate
Petitpierre et al., 2015 [104]	73 (on dialysis)	76	Volume reduction, lack of space for kidney allograft	6 (7.9)	2.97 ± 1.56	15 (18.3)	0 (0)	Not reported
Suwabe et al., 2016 [105]	449 (on dialysis)	844 (14 unilateral REA)	Volume reduction, volume-related complaints	89 (19.8)	Not reported	'Most'	Not reported (the text cites 13 patients)	3.34% (15 total deaths, 3 RAE-related)
Sakuhara et al., 2015 [108]	15 (on HD)	30	Volume-related complaints	1 (6.7)	8 [8–13]	'All'	1 (6.7)	6.7% (unknown whether this was RAE-related)
Suwabe et al., 2017 [110]	188 (on HD)	376	Quality of life, volume-related complaints	Not reported	10	'All'	Not reported (the text cites 1 patient)	0.5%
Pierre et al., 2020 [106]	21 (on PD)	21	Volume reduction, lack of space for kidney allograft	6 (28.6)	5.0 [4.0–6.0]	Not reported	Not reported	Not reported
Del Tatto et al., 2022 [107]	29 (9 PD and 20 HD)	31	Volume reduction, lack of space for kidney allograft	1 (3.4)	3.8 (3–6)	10 (32.2)	0 (0)	Not reported, 30-day mortality 0%

Data are expressed as mean ± SD, median [interquartile range], or median (range). HD, hemodialysis; PD, peritoneal dialysis; RAE, renal artery embolization.

for temporary or permanent transition to hemodialysis [106]. This was likely the result of the minimally invasive nature of RAE, which can be performed with a single puncture to reach the femoral artery and under local anesthesia, although it has also been described to be performed under general anesthesia [106, 107]. Given that RAE can be performed under local anesthesia, RAE might be preferred when a patient has a high risk of complications related to general anesthesia and surgery during native nephrectomy. A retrospective study from 2020 also found a shorter hospital stay of 5 days in patients treated with RAE compared with 8.5 days in those who underwent open native nephrectomy [106]. Other studies report an average duration of hospital stay between 3 and 10 days after RAE [104–108].

In some hospitals, RAE is currently the first-line treatment in ADPKD patients with massively enlarged kidneys, with the aim of facilitating implantation of a kidney graft in the iliac fossa [111]. However, a disadvantage of the procedure is that the effect is not immediate, because a significant volume reduction is achieved only after 3–6 months, which may translate into a temporary contraindication for transplantation [104]. Some patient characteristics have been related to greater success of RAE, such as younger patients, shorter time on dialysis or with associated hypertension [105]. When embolization is not successful, nephrectomy may still be possible in cases of persistent pain or insufficient volume reduction [104, 106, 107]. The literature varies on the difficulty of surgical nephrectomy post-embolization. Some argue that surgery is more complicated due to fibrosis post-embolization, while others found nephrectomy facilitated by volume and renal blood flow reduction [104].

There are no prospective studies that analyze the complications of RAE and compare them with a nephrectomy group. Major complications have been reported (0%–5%), among which are intestinal perforation due to obstruction after RAE, cyst infection, gastrointestinal bleeding in some patients using non-steroidal anti-inflammatory agents for analgesia after the procedure, pseudoaneurysm at the femoral artery puncture site, thrombotic complications, and others [104–107, 110]. RAE can also result in a post-embolization syndrome with severe pain, fever, nausea and vomiting caused by necrosis of kidney tissue. The occurrence rate of this syndrome has been described to range between 30% and 100% [104, 105, 107, 108, 110]. To mitigate complaints related to this post-embolization syndrome, patients should be prescribed sufficient analgesics and moderate sedation may be added [103].

Mortality rate of RAE has been described to be low in most publications, although in one study 1-year mortality rate after embolization was 3.3% [104–107, 110]. However, this was a retrospective study in 449 patients with ADPKD undergoing bilateral RAE, without a control group, in which only 3 out of 15 deaths were attributed to the procedure. Nonetheless, together with the large differences in reported rates of post-embolization syndrome, this underlines that there are differences in the results of RAE between centers.

Taken together, there is currently insufficient evidence to recommend RAE over nephrectomy, but the aforementioned preliminary results indicate that RAE may be an alternative to nephrectomy for certain indications (volume reduction, cyst hemorrhage, but not cyst infections of malignancy). For these indications, it can be considered after multidisciplinary consultation, weighing the risks related to anesthesia and surgery associated with native nephrectomy against the lower success chance and risk of post-embolization syndrome associated with RAE, as well as experience at the center, patient condition and preference, and dialysis mode.

Practice point 11: Renal artery embolization may be an alternative to surgical nephrectomy and can be considered after multidisciplinary consultation, weighing the risks related to anesthesia and surgery associated with native nephrectomy against the lower success chance and risk of post-embolization syndrome associated with renal embolization. Experience at the center, patient condition and preference, and dialysis mode should be taken into account.

STRENGTHS AND LIMITATIONS

Several reviews have been published about nephrectomy in ADPKD [112–115]. A recent KDIGO Guideline also mentions this topic [16]. The present systematic review, however, represents the most comprehensive overview of all aspects concerning nephrectomy in patients with ADPKD to date, including more in-depth surgical considerations. The results of our review are in large part based on objective findings obtained via a systematic literature review, summarized in formal meta-analyses using standard software when possible. These findings were complemented with data obtained from a questionnaire distributed among representatives of the three different medical specialties that are involved with nephrectomies in this patient category (nephrologists, urologists, and transplant surgeons). The findings of this process were discussed with the European Renal Association Working Group Genes & Kidney, specific experts invited by the Working Group, the European Association of Urology Section of Transplantation Urology, and patient representatives of PKD International. Expert consensus concerning the developed practice points was reached through a Delphi method. Thus, based on the above-mentioned considerations, the present review is the most complete and multidisciplinary publication concerning this topic. This study also has limitations, the main one being that all included studies are retrospective case series, which may be subject to selection bias. However, the risk of bias was determined to be low in almost all included publications when using a dedicated scoring system. If heterogeneity was found, sensitivity analyses were performed, identifying no major causes of bias explaining the variability in results. Another limitation is that not all publications reported all variables of interest. However, in most cases this information was kindly provided by the corresponding authors upon request, thus enhancing the generalizability and comprehensiveness of the analyses.

CONCLUSIONS

This consensus statement gives a comprehensive overview of important clinical questions concerning nephrectomy in patients with ADPKD. Based on the findings of the systematic literature search and the exploratory questionnaire, we propose a dedicated workup to decide whether a nephrectomy is indicated in the period just before a kidney transplantation. To assess the size of the polycystic kidneys, we suggest performing an MRI or CT. This will give an indication of whether there is lack of space for a kidney allograft and whether the polycystic kidneys (and liver) may be the cause of existing gastrointestinal complaints. The patient perspective should be explored using the ADPKD-PDS and PLD-Q questionnaires containing questions about the severity of complaints, which can be filled out by patients before consultation. After a multidisciplinary evaluation, the possible indications, benefits and risks of a nephrectomy should be discussed with the patient to allow making a shared decision. When there is insufficient space for a kidney allograft, we suggest performing a unilateral

procedure just before or combined with kidney transplantation. The choice of which surgical approach to pursue is dependent on the availability of (robot-assisted) laparoscopy, the surgeon's experience, and the size of the polycystic kidneys. Nephrectomy after transplantation is, in general, described to be just as safe as before transplantation, indicating that there seems to be no need to perform pre-emptive nephrectomy to avoid surgery after transplantation. Future research is needed to evaluate the role of renal artery embolization as an alternative to nephrectomy, as well as to establish score thresholds for pain and gastrointestinal symptom questionnaires that would warrant consideration of nephrectomy. This will help improve targeted decision-making processes in the future. In addition, the effect of nephrectomy on dyspnea and patient self-image, and the required abdominal space for successful kidney transplantation should be investigated in future studies.

SUPPLEMENTARY DATA

Supplementary data are available at [Nephrology Dialysis Transplantation](#) online.

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CONFLICT OF INTEREST STATEMENT

All authors stated that they did not have conflicts of interest for this specific article.

DATA AVAILABILITY STATEMENT

The data underlying this article will be shared upon reasonable request to the corresponding author.

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