

Simultaneous Right-Sided Nephrectomy with Orthotopic Liver/Kidney Transplantation - An Alternative Method for Patients with Autosomal Dominant Polycystic Liver and Kidney Disease

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Research article

Keywords: kidney transplantation, liver transplantation, autosomal dominant polycystic liver and kidney disease

Posted Date: April 28th, 2020

DOI: <https://doi.org/10.21203/rs.3.rs-21666/v1>

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Version of Record: A version of this preprint was published at Langenbeck's Archives of Surgery on May 26th, 2021. See the published version at <https://doi.org/10.1007/s00423-021-02206-9>.

Abstract

Background: In patients suffering from autosomal dominant polycystic liver and kidney disease (ADPLKD), organ transplantation often poses a technical challenge due to the large volume of both organs. To simplify the transplantation procedure by improving the exposure of anatomical structures, we introduce a novel surgical technique.

Methods: Seven patients with ADPLKD were evaluated for simultaneous liver and kidney transplantation at the Jena University Hospital. The modified transplantation approach included hepatectomy and right-sided nephrectomy. After reperfusion of the liver graft, orthotopic kidney transplantation to the right-sided hemiabdomen was performed.

Results: Preoperatively, the median liver volume and right kidney volume were morphologically determined by CT to be 9343 ml (\pm 3698.56) and 2770 ml (\pm 1128.78). The mean operation time was 342.43 minutes (\pm 68.77). Postoperatively, the patients were treated 6.28 days (\pm 2.50) in the intensive care unit and were discharged from the surgical ward approximately 28 days (\pm 5.66) after the operation with normal graft function.

Conclusion: The modified simultaneous liver and kidney transplantation technique is an effective alternative surgical transplantation method for patients with ADPLKD. The new transplantation technique simplifies the transplantation procedure by improving the exposure of anatomical structures.

Background

Since first reported by Margreiter et al. in 1984 (1), the simultaneous transplantation of the liver and kidney transplantation has been accepted as an effective surgical method in clinical practice, having a good patient outcome (2) and significantly improving the quality of life (3).

One indication for simultaneous organ transplantation is autosomal dominant polycystic liver and kidney disease (ADPLKD) (Potter III). In the presence of ADPLKD, kidney function can remain normal for a long time despite massive enlargement of the organ caused by the cysts (4). In 34% to 78% of patients with adult polycystic kidney disease, multiple liver cysts exist simultaneously (5) (6). The overall liver function in these patients is often maintained, but they may suffer from a number of nonspecific clinical symptoms, such as postprandial flank or abdominal pain, chronic nausea and vomiting, or shortness of breath. These symptoms often correlate with the size or position of the polycystic organs. Complications such as renal anemia, arterial/renal hypertension, cyst infection, significant weight loss, ureter or bile duct obstruction, nephrolithiasis, or hepatic venous-outflow obstruction can occur with disease progression.

The therapeutic procedure for ADPLKD is usually aligned to Gigot's classification (10). For type II and III ADPLKD patients, simultaneous liver and kidney transplantation provides the only curative treatment option (9) (11). In 42% of all patients with ADPLKD, simultaneous liver and kidney transplantation is necessary (12). In general, the simultaneous transplantation of these organs is a very rare procedure in the EUROTRANSPLANT region. Only forty-five combined transplantations were performed in 2017 (18), but only 15% of these were indicated for ADPLKD (19).

Generally, the clinical standard transplantation procedure is based on two steps: 1.) the orthotopic liver transplantation, and 2.) the heterotopic kidney transplantation in the iliac fossa via a second surgical incision (two-step cLKTx). Currently, the surgical technique of simultaneous liver and kidney transplantation is subject to individual variability (13) and a center-specific approach (14) (15). The challenge in all existing surgical techniques is to reduce the clinical symptoms resulting from increased polycystic organ volume and to simplify the transplantation procedure by improving the representation, scope, and details of the anatomical structure.

In this article, we present the feasibility of a newly modified surgical technique for orthotopic simultaneous liver and kidney transplantation in patients with ADPLKD. Furthermore, we describe the three-month follow-up of patients with ADPLKD who have been transplanted with this modified technique as our standard since September 2016.

Methods

Preoperative clinical parameters:

The modified surgical technique of simultaneous liver and kidney transplantation has been used at the Transplantation Centre of the Jena University Hospital, Germany, since September 2016 as the standard operation technique for patients with ADPLKD. The respective patients underwent a comprehensive evaluation of possible contraindications. Furthermore, the internal interdisciplinary transplantation committee approved the indication for simultaneous organ transplantation prior to listing. The preoperative clinical parameters of the patients are derived from the internal hospital data reporting system as well as from the data transmitted to EUROTRANSPLANT. The characteristics of the donor and the recipient, along with the results of clinical routine examinations, were evaluated in the reported data analysis (ethical committee application number: 4428-05/15).

Technique of the modified simultaneous liver and kidney transplantation:

The abdominal cavity was opened up from the xiphoid down to the anterior superior iliac spine as a right-sided L-incision. The abdominal cavity was exposed by bilateral retraction of the ribcage to obtain access to the liver and the right kidney. Due to the increased volumes of the liver and the kidney, the preparation of the anatomical structures, especially the retrohepatic vena cava, can prove to be difficult. To simplify the procedure, individual cysts were opened during the preparation.

The right-sided nephrectomy of the polycystic altered organ was performed first to facilitate the hepatectomy, to simplify the transplantation procedure and to reduce the intra-abdominal volume. In preparation for this step, the right-sided colon and the duodenum were mobilized, and the renal vein and artery were identified and dissected. During nephrectomy, special attention must be paid to maintaining a long ureter with a surrounding mass of vessel-supplied tissue.

Next, the liver was mobilized from the ligamentary structures and prepared for hepatectomy, including the retrohepatic vena cava. Before hepatectomy, the liver and kidney graft were prepared at the back table for the implantation.

Subsequently, the liver transplantation followed its usual course with the caval replacement technique and without the use of a temporary portocaval shunt or veno-venous bypass. Reperfusion was performed simultaneously, both portal and arterial. The bile duct was reconstructed as duct-to-duct modified end-end anastomoses.

After the completion of the liver transplantation, kidney transplantation in the orthotopic retroperitoneal position was performed via the pre-established right-sided L-incision. The venous anastomosis was made directly to the inferior V. cava below the original stump of the right renal vein. Arterial reconstruction was established for the right common iliac artery of the recipient using an arterial interponate of the donor iliac artery, guided retroperitoneally for this means. After the reperfusion of the kidney graft and the preparation of the recipient ureter, an end-end uretero-ureterostomy was performed with a double-J catheter splinting. The established L-incision was closed after inserting drainage tubes (two 20-gauge French tubes placed subhepatically and subphrenically).

CT-scan and ultrasound imaging:

As part of the routine evaluation procedure, a full-body computer tomography (CT-scan; CT Revolution 256 slice, GE Healthcare, USA) was performed for each potential recipient. The organ volume of the polycystic liver and kidney was calculated using Cerner SkyVue® Distribution (Version 2014.01.05, Cerner, USA) and the available CT-scan results. To visualize the transplanted organs and their vascular supply, clinically indicated postoperative CT-scans were reconstructed with MeVis® (Fraunhofer MeVis, Lübeck, Germany) (Fig. 1). To evaluate the postoperative course, the recipients underwent routine ultrasound examinations (Vivid S70, GE Healthcare, USA). During routine ultrasound exams, the perfusion of the transplanted organs was examined by determining the arterial and venous blood velocity and arterial vascular resistance.

Results

Preoperative clinical parameters:

Since 2016, the modified simultaneous liver and kidney transplantation technique has been the standard operation technique for patients suffering from ADPLKD at the Transplantation Centre of Jena University Hospital. Until October 2018, a total of seven patients were treated using this modified technique.

At the time of transplantation (Table 1), the patients were on average 54.14 years (\pm 3.98) old. Four of them were female. All patients had restricted liver function with a laboratory Model of End-Stage Liver Disease score (lab-MELD) of 18.57 points (\pm 2.37). All patients fulfilled the standard exceptional criteria for prioritization on the waiting list according to the EUROTRANSPLANT manual (standard exceptional Model of

End-Stage Liver Disease (SE-MELD): 29.29 points (± 2.06) (16). The average waiting time prior to the transplantation was 834.57 days (± 832.23). The patients were categorized as ASA-score III with comorbidities such as normochromic normocytic anemia, renal hypertension, or type 2 diabetes. The mean body mass index (BMI) of the patients was 27.09 kg/m² (± 5.59). During the routine evaluation procedure, morphological analysis through a CT-scan revealed an average liver volume of 9343 ml (± 3698.56) and an average right kidney volume of 2770 ml (± 1128.78) (Table 2). All patients were categorized as type III according to Gigot’s ADPLKD classification (Figs. 2 and 3). Donor-specific parameters are presented in Table 2.

Table 1
Preoperative clinical parameters of patients with ADPLKD evaluated for modified liver and kidney transplantation

Patient-ID	P-001	P-002	P-003	P-004	P-005	P-006	P-007	Average (SD)
Age (y)	52	50	57	54	61	55	50	54.14 (± 3.98)
Waiting time (d)	781	589	1032	2602	252	250	336	834.57 (± 832.23)
Gender	male	female	female	male	female	female	male	-
Preoperative dialysis	no	no	yes	no	no	no	no	-
Body weight (kg)	77	53	73	117	73	83	84	80 (± 19.28)
Body height (m)	1.83	1.56	1.68	1.79	1.62	1.60	1.95	1.72 (± 0.14)
BMI (kg/m ²)	23.10	21.80	25.90	36.50	27.82	32.42	22.09	27.09 (± 5.59)
Blood Typ (*)	0 (-)	0 (+)	0 (+)	A (+)	0 (+)	0 (+)	0 (+)	-
Lab-MELD	17	15	20	19	20	17	22	18.57 (± 2.37)
SE-MELD	29	33	29	29	26	29	30	29.29 (± 2.06)
Values are presented in average and standard deviation (SD); * Rhesus-factor; BMI: Body mass index; Lab-MELD: laboratory-model of end stage liver disease; SE-Meld: standard exceptional-model of end stage liver disease								

Table 2
Donor-, intraoperative- and organ-specific parameters in patients with modified liver and kidney transplantation

	Patient-ID	P-001	P-002	P-003	P-004	P-005	P-006	P-007	Average (SD)
Donor-specific parameter	Age	55	79	61	72	63	61	51	-
	Gender (M/F)	M	F	F	F	F	F	F	-
	Body weight (kg)	90	60	63	80	70	72	60	-
	Body height (m)	1.90	1.60	1.70	1.65	1.70	1.67	1.60	-
	BMI (kg/m ²)	24.90	23.40	21.80	29.40	24.20	25.80	23.40	-
	Extended donor	yes ⁽¹⁾	yes ^(2,3)	yes ^(1,3)	yes ^(1,2)	yes ^(3,4,5)	yes ⁽³⁾	yes ⁽⁵⁾	-
Intraoperative parameter	Operation time (min)	326	236	288	413	322	419	393	342.43 (± 68.77)
	CIT Liver (min)	420	516	535	497	395	621	208	456 (± 132.46)
	WIT Liver (min)	25	31	32	29	32	56	46	35.86 (± 11.01)
	CIT Kidney (min)	559	563	648	457	488	741	550	572.29 (± 95.05)
	WIT Kidney (min)	12	10	21	10	20	21	34	18.29 (± 8.58)
Organ-specific parameter	Liver vol. (ml)	11300	7394	6263	9867	10243	4576	15758	9343 (± 3698.56)
	Kidney vol. rig. (ml)	2931	1899	1408	4336	3881	1714	3221	2770 (± 1128.78)
	Liver wt (g)	4500	3600	4220	4960	5585	1775	8210	4692.86 (± 1966.66)

Values are presented in average and standard deviation (SD); CT-volumetry using CT Revolution 256 slice, GE Healthcare, USA; Cerner SkyVue® Distribution, version 2014.01.05, Cerner, USA; M: male, F: female; BMI: Body mass index; wt: weight; vol: volume; Extended donor criteria reason: 1.: non-traumatic subarachnoid haemorrhage, 2.: donor age, 3.: ICU stay more than 7 days; 4.: Cancer in medical history; 5.: Limited graft function

Patient-ID	P-001	P-002	P-003	P-004	P-005	P-006	P-007	Average (SD)
Kidney wt (g)	2000	1028	1900	3860	2908	984	1635	2045 (\pm 1032)
Values are presented in average and standard deviation (SD); CT-volumetry using CT Revolution 256 slice, GE Healthcare, USA; Cerner SkyVue® Distribution, version 2014.01.05, Cerner, USA; M: male, F: female; BMI: Body mass index; wt: weight; vol: volume; Extended donor criteria reason: 1.: non-traumatic subarachnoid haemorrhage, 2.: donor age, 3.: ICU stay more than 7 days; 4.: Cancer in medical history; 5.: Limited graft function								

Intraoperative evaluation of simultaneous liver and kidney transplantation

The mean operation time (Table 2) of the modified combined liver and kidney transplantation was 342.43 minutes (\pm 68.77). The establishment of the vascular liver (WIT liver) and kidney (WIT kidney) anastomoses took 35.86 minutes (\pm 11.01) and 18.29 minutes (\pm 8.58), respectively.

On average, 785.71 ml (\pm 393.40) of human albumin was infused to compensate for the intravascular volume deficiency. In only two patients was it necessary to compensate for the coagulopathy ROTEM driven by the transfusion of fresh frozen plasma (FFP).

Postoperative evaluation of simultaneous liver and kidney transplantation:

After leaving the operation theater, the patients were observed in the ICU for approximately 6.28 days (\pm 2.50). Immediate extubation and hemodynamic stabilization were achieved in all patients.

In only one case did delayed kidney function occur. The initially impaired renal function in this patient (P-002) resulted in hypervolemia with subsequent respiratory insufficiency. After reintubation and one-off hemodialysis (Table 3; Clavien-Dindo classification IVa), the kidney graft function stabilized promptly, and the patient could be transferred to the surgical ward on POD 11. During hospitalization, more than half of the patients had a minor complication, which was classified as type II according to the Clavien-Dindo classification.

Table 3

Preoperative, postoperative and three-month follow-up parameters in patients with modified liver and kidney transplantation

	Patient-ID	P-001	P-002	P-003	P-004	P-005	P-006	P-007
Preoperative	CREA ($\mu\text{mol/l}$)	277	257	531	356	334	360	542
	eGFR (ml/min)	21.17	18.1	7.1	15.8	12.2	11.7	9.8
	UREA ($\mu\text{mol/l}$)	17.1	15.9	14.7	4.7	20.6	18	21.8
Postoperative follow-up	postoperative Dialysis (n)	no	yes (1)	no	no	no	no	no
	PRC (n)	1	6	2	0	1	0	6
	FFP (n)	0	0	0	0	0	0	0
	Albumin (ml)	0	300	0	1000	500	0	650
	Urine secretion POD 1 (ml)	7010	3140	5160	5600	780	2800	1500
	Urine secretion POD 2 (ml)	3850	1800	2620	4100	6920	4200	3140
	Urine secretion POD 3 (ml)	1430	990	2390	2900	1440	3850	2010
	ICU stay (d)	4	11	8	4	5	6	6
	Hospitalisation (d)	27	28	37	33	21	22	28
	Clavien-Dindo	II	IVa	IIIa	II	II	IIIa	II
Three month follow-up	AST ($\mu\text{mol/l}$)	0.33	0.87	0.58	0.37	0.26	0.32	0.18
	ALT ($\mu\text{mol/l}$)	0.43	1.12	0.87	0.36	0.19	0.33	0.10
	Ap ($\mu\text{mol/l}$)	1.29	2.21	1.25	0.66	1.15	1.59	1.36
	GGT ($\mu\text{mol/l}$)	0.55	2.84	0.62	0.44	0.39	2.34	0.5
	Lip ($\mu\text{mol/l}$)	0.27	0.24	0.38	0.84	0.55	1.09	0.26
	CREA ($\mu\text{mol/l}$)	112	134	73	148	83	141	164
	eGFR (ml/min)	64.7	39.8	79.1	45.6	65.9	35.9	41.2
	UREA ($\mu\text{mol/l}$)	9.2	5.1	3.5	10.7	6.3	8.3	8.9
	ALB (g/l)	39	42	34	35	38	36	32

AST: Aspartat-Aminotransferase, ALT: Alanin-Aminotransferase, AP: Alkaline phosphatase, GGT: γ -Glutamyltransferase, Lip: Lipase, CREA: Creatinine, eGFR: estimated glomerular filtration rate, UREA: urea, ALB: Albumin, INR: international Normalized Ratio; BIL: bilirubin, ChE: Cholinesterase

PRC: packed red cells, FFP: fresh frozen plasma; POD: postoperative day

Patient-ID	P-001	P-002	P-003	P-004	P-005	P-006	P-007
INR	1	0.9	1	1	1	0.9	1.1
BILI (μmol/l)	6	24	9	6	14	4	7
ChE (μmol/l)	150	121	161	124	117	137	73
AST: Aspartat-Aminotransferase, ALT: Alanin-Aminotransferase, AP: Alkaline phosphatase, GGT: γ-Glutamyltransferase, Lip: Lipase, CREA: Creatinine, eGFR: estimated glomerular filtration rate, UREA: urea, ALB: Albumin, INR: international Normalized Ratio; BILI: bilirubin, ChE: Cholinesterase							
PRC: packed red cells, FFP: fresh frozen plasma; POD: postoperative day							

The graft function in the other six patients normalized promptly, and no irregularities occurred in the remaining clinical course of these patients. The routine ultrasound examination showed regular postoperative results with normal perfusion parameters and no signs of cholestasis or urinary retention (Table 4). There was no relevant postoperative bleeding. The immunosuppressive rules and standards based on tacrolimus (target level 8–12 ng/ml within the first 3 months), mycophenolate, and steroids were gradually tapered, and were discontinued, depending on the patient, over a period of 6 months after transplantation. Basiliximab (20 mg at POD 0 and POD 4) was used for the immunosuppressive induction. Following the removal of the double-J catheter, patients were discharged after 28 days (\pm 5.66) with normal liver and kidney function (data not shown). Furthermore, there was no manifestation of bile leakage, ureter stenosis, or incisional hernia during the postoperative observation period.

Table 4
 Sonographic findings using a Vivid S70 (GE Healthcare, USA) in the first three postoperative days in patients with modified liver and kidney transplantation

	Parameters	Average (SD)
POD 1	PVF (cm/s)	19.14 (± 8.95)
	HAF (cm/s)	59.67 (± 39.02)
	RI-HA	0.55 (± 0.13)
	RVF (cm/s)	25.55 (± 28.35)
	RAF (cm/s)	39.17 (± 13.04)
	RI-RA	0.63 (± 0.07)
POD 2	PVF (cm/s)	28.33 (± 10.56)
	HAF (cm/s)	44.57 (± 13.64)
	RI-HA	0.58 (± 0.11)
	RVF (cm/s)	12.5 (± 0.71)
	RAF (cm/s)	26.33 (± 8.14)
	RI-RA	0.63 (± 0.06)
POD 3	PVF (cm/s)	29.52 (± 16.81)
	HAF (cm/s)	41 (± 19.36)
	RI-HA	0.62 (± 0.08)
	RVF (cm/s)	14 (± 4.24)
	RAF (cm/s)	25.42 (± 6.47)
	RI-RA	0.58 (± 0.05)
Values are presented in average and standard deviation (SD); PVF: portal vein flow, HAF: hepatic artery flow, RI-HA: Resistance-Index hepatic artery, RVF: renal vein flow, RAF: renal artery flow, RI-RA: Resistance-Index renal artery, POD: postoperative day		

The postoperative pathological examination (Table 2) confirmed the preoperative calculated liver and kidney dimensions (mean liver weight 4692.86 g (± 1966.66); mean kidney weight 2045 g (± 1032.78)). Opening single cysts during organ explantation is responsible for the difference between the pre- and postoperative examination.

The three-month follow-up confirmed excellent graft function (Table 3) with normal liver synthesis and normal kidney function (retention parameters) in all patients. The indication for additional left-sided nephrectomy was not given for any patient during the follow-up period.

Discussion

The treatment of patients with ADPLKD is always individual. The respective strategies are oriented to the patients' individual symptoms as well as the patient- and organ-specific comorbidities. Controlling the symptoms associated with the cysts and preventing disease-specific complications are the primary aims of any therapy. In up to 90% (17) of all patients with ADPLKD, radiological or surgical interventions such as ethanol injection or cyst fenestration are the preferred therapy. By using these strategies, a transient reduction in symptoms can be achieved in approximately 92% of all patients (8) (9). Unfortunately, in 24% of these patients, the cysts reoccur. Depending on the therapy received, a morbidity rate of 23–50% and a mortality rate of 2–3% can be assumed (8) (9).

In patients with progressive symptoms and resulting comorbidities, transplantation remains the only curative treatment option. Transplantation can decrease the mortality and morbidity associated with the comorbidities and improve the quality of life (2) (3). Simultaneous liver and kidney transplantation, due to the massive cystic enlargement seen in ADPLKD, is technically challenging and varies depending on patients (13) and center-specific (14) (15) factors. This article demonstrates the secure feasibility of a new technique based on simultaneous orthotopic liver and kidney transplantation and right nephrectomy through a single incision in a sample of patients with ADPLKD.

Previously, Jochmans and colleagues (20) also described simultaneous organ transplantation through an individual median laparotomy. The kidney graft was placed in a retroperitoneal heterotopic position in the iliac fossa in a peritoneal pocket that was formed by peeling down the peritoneum from the midline (one-step cLKTx). Jochmans et al. documented a similar mortality and morbidity in the one-step cLKTx and the two-step cLKTx. Furthermore, the operation time (one-stage cLKTx 6.8 h (4.1 h-9.3 h) vs. two-step cLKTx 9.0 h (8.7 h-10.1 h)), as well as the cold ischemic time (one-step cLKTx 8.1 h (6.4 h-9.3 h)), could be reduced using the modified technique (20).

The novelty of our modified technique is right-sided nephrectomy during combined transplantation. Compared to Jochmans et al., our modified liver and kidney transplantation procedure could reduce the operation time by more than one hour (one-stage cLKTx 6.8 h vs. orthotopic simultaneous liver and kidney transplantation 5.45 h). This reduction is due to the right-sided nephrectomy. In addition, the right-sided nephrectomy facilitates the subsequent liver transplantation and the orthotopic transplantation of the kidney through the already prepared retroperitoneum. This surgical strategy results in a reduced intra-abdominal organ volume and improves the representation and detailed view of the anatomical structures during the preparation for hepatectomy and the implantation of the liver graft. Additionally, the vascular structures are openly accessible and can be prepared for subsequent kidney transplantation. Furthermore, the scar surface is reduced, since simultaneous transplantation can be carried out through a single incision.

Apart from the advantages of the modified transplantation technique, the morbidity and mortality rates are comparable with all established simultaneous transplantation techniques. Common perioperative complications after simultaneous transplantation are delayed graft function (18%), infections (24%) and cardiovascular complications (11%) (23). The evaluation of the one-stage cLKTx of Jochmans et al. showed a complication rate of up to 25% according to the Clavien-Dindo classification \geq III. This rate is similar to that

of the classical transplantation method (20) as well as our modified simultaneous liver and kidney transplantation. Furthermore, surgical complications associated with our modification of the surgical technique were not observed. Except in one case, conservative therapeutic management successfully treated all complications. Additionally, frequent complications of heterotopic kidney transplantation, such as lymphocele, could be excluded by orthotopic transplantation of the kidney in the physiological position. Furthermore, the modified simultaneous organ transplantation reduced the ICU and in-hospital time of patients with ADPLKD. The patients spent, on average, 6.28 days (± 2.50) in the ICU and 28 days (± 5.66) in the hospital following the transplantation procedure. In contrast, patients who were transplanted using the standard transplantation technique spent an average of 9 days (± 8.4) in the ICU and in total 33 days (± 9.7) in the hospital (13).

The limitation for applying this modified transplantation technique is the obligatory nephrectomy. Each patient should be individually evaluated as to whether a simultaneous orthotopic liver and kidney transplantation with accompanying right-sided nephrectomy is the therapy of choice, which depends on the clinical symptoms of the patient and the intraoperative course. The median nephrectomy rate ranged from 33% (unilateral) to 67% (bilateral) (20) for all patients with ADPLKD, independent of the respective therapy strategy. Some transplant centers recommend a total nephrectomy before or during the transplantation in this patient group (21). However, simultaneous orthotopic transplantation with accompanying right-sided nephrectomy offers clear advantages over a two-sided procedure by sparing one operation, including narcosis, as well as improving the operative access during the transplantation procedure. In contrast to the indication for nephrectomy, there is less variability in the procedure of orthotopic liver transplantation. Usually, liver transplantation is performed either in vena cava interposition or using the “piggy-back” technique. Studies have consistently shown that there is no significant difference in graft or patient survival between the two techniques. Generally, the 1-year and 5-year survival rates after simultaneous liver and kidney transplantation are 88% and 80%, respectively (22) (2) (3).

The interpretation of the results of the study is subject to further limitations. The small size of our study is one restriction and is due to the relatively rare procedure of simultaneous liver and kidney in the EUROTRANSPLANT region and the relatively rare prevalence of ADPLKD in the general population. Furthermore, simultaneous organ transplantation is considered in patients with type II and III ADPLKD (9) (11) according to Gigot’s classification. Another restriction of the study is the currently missing long-term course of our patients treated with the modified orthotopic liver and kidney transplantation. However, these results are important to provide a comprehensive assessment of the modified technique.

In summary, the modified orthotopic liver and kidney transplantation technique is feasible and promising for patients suffering from polycystic liver and renal degeneration. The procedure yields a comparable mortality and morbidity rate to the classical transplantation method (13) (20). We were also able to demonstrate that our surgical technique reduced the relevant operation-specific parameters (operation time and warm ischemia time) compared to studies conducted using other techniques. Further experience and data are needed, especially in long-term outcomes, to clearly evaluate our promising results. Additionally, long-term surgical complications such as incisional hernia and vascular, bile duct or urinary complications are subjects for further research. Furthermore, the described modified surgical technique must be compared to the already

established standard simultaneous orthotopic liver and heterotopic kidney transplantation. A prospective randomized clinical trial is planned to compare the modified orthotopic surgical technique with the clinical standard heterotopic operation procedure. These results will be used to consider broadening the application of the modified orthotopic surgical technique to other indications for combined transplantation in patients with end-stage liver and renal disease.

Conclusion

In conclusion, the modified orthotopic liver and kidney transplantation technique is an alternative technique for combined liver and kidney transplantation. The transplantation technique simplifies the transplantation procedure without negatively influencing the outcome of the patient.

Abbreviations

ADPLKD: Autosomal dominant polycystic liver and kidney disease

PDK-1: Pyruvate dehydrogenase kinase 1

PDK-2: Pyruvate dehydrogenase kinase 2

Two-step cLKTx: Standard simultaneous liver and kidney transplantation

Art: Arteria

V: Vena

Lig: Ligamentum

ICU: Intensive care unit

Lab-MELD: Laboratory Model of End-Stage Liver Disease

SE-MELD: Standard exceptional Model of End-Stage Liver Disease

BMI: Body mass index

CT-scan: Computed tomography

CRC: Concentrated red cells

FFP: Fresh frozen plasma

CIT: Cold ischemic time

WIT Warm ischemic time

PVF: Portal vein flow

HAF: Hepatic artery flow

RI-HA: Resistance-Index hepatic artery

RVF: Renal vein flow

RAF: Renal artery flow

RI-RA: Resistance index-renal artery

POD: Postoperative day

AST: Aspartate aminotransferase

ALT: Alanine aminotransferase

AP: Alkaline phosphatase

GGT: γ -Glutamyltransferase

Lip: Lipase

CREA: Creatinine

eGFR: Estimated glomerular filtration rate

UREA: Urea

ALB: Albumin

INR: International Normalized Ratio

BILI: Bilirubin

ChE: Cholinesterase

One-step cLKTx: Modified simultaneous liver and kidney transplantation according to Jochmans et al.

Declarations

Ethics approval and consent to participate

This retrospective database analysis study was approved by the institutional review board of University Hospital Jena Ethics Committee (ethical committee application number: 4428-05/15), and the requirement to obtain informed consent was waived.

Consent for publication

All authors consent to the publication.

Availability of data and material

The datasets used and analyzed during the current study are available from the corresponding author on reasonable request.

Competing interests

The authors declare no competing interests.

Funding

This work was supported by funding of the Foundation “Else Kröner-Fresenius-Stiftung”.

Authors' contributions

All Authors read and approved the manuscript

PF designed the study, analyzed data and wrote the manuscript.

HMT analyzed data and wrote the manuscript

SL collected data and analyzed data.

FD collected data and analyzed data.

RA collected data and analyzed data.

FR collected data analyzed data and edited the manuscript

US designed the study, analyzed data and edited the manuscript.

Acknowledgements

Not applicable

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Figures

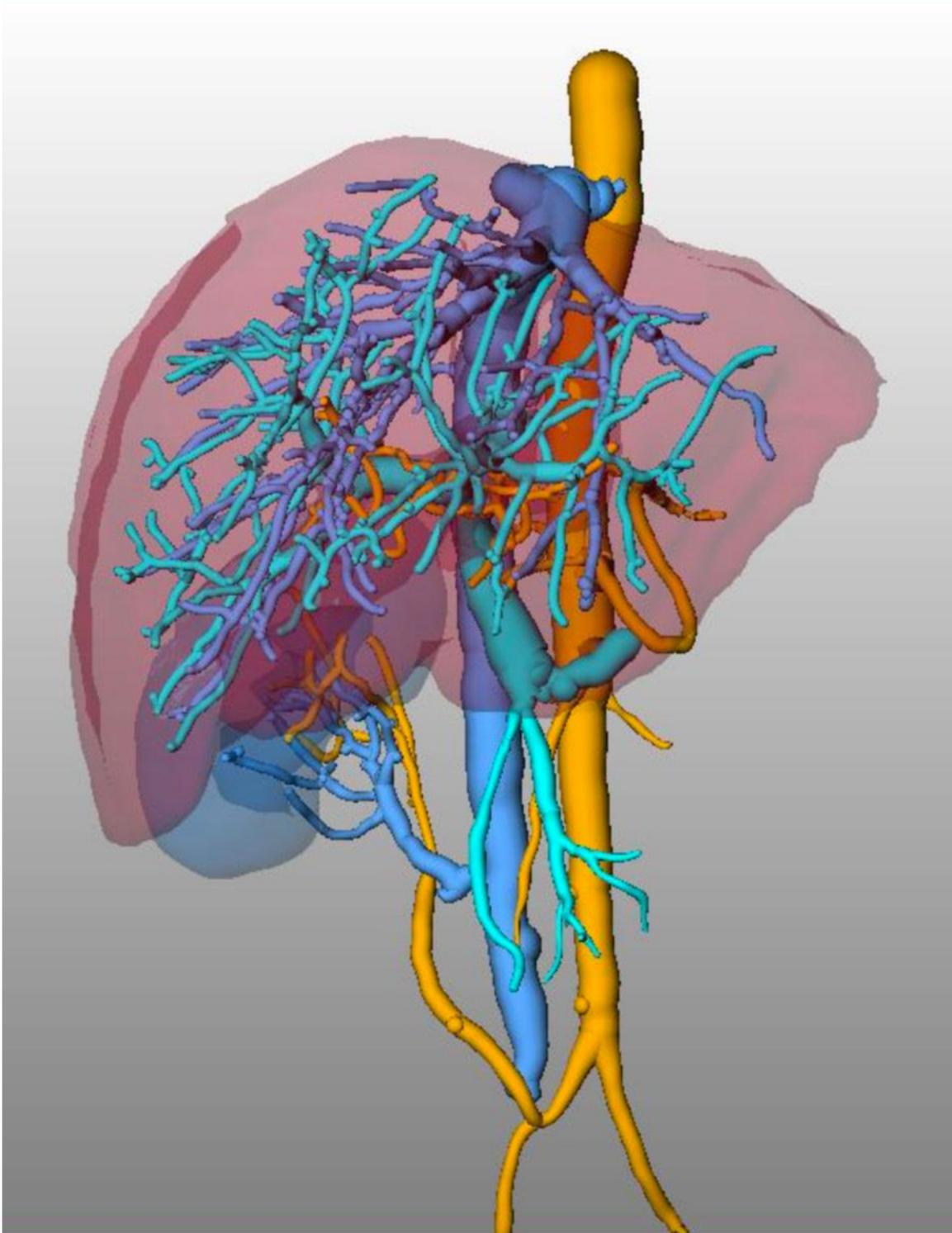


Figure 1

CT reconstruction (MeVis® (Fraunhofer MeVis, Lübeck, Germany)) after the transplantation of a patient with polycystic liver and kidney degeneration. Illustration of the transplant organs with arterial, portal, and venous vascular anatomy.

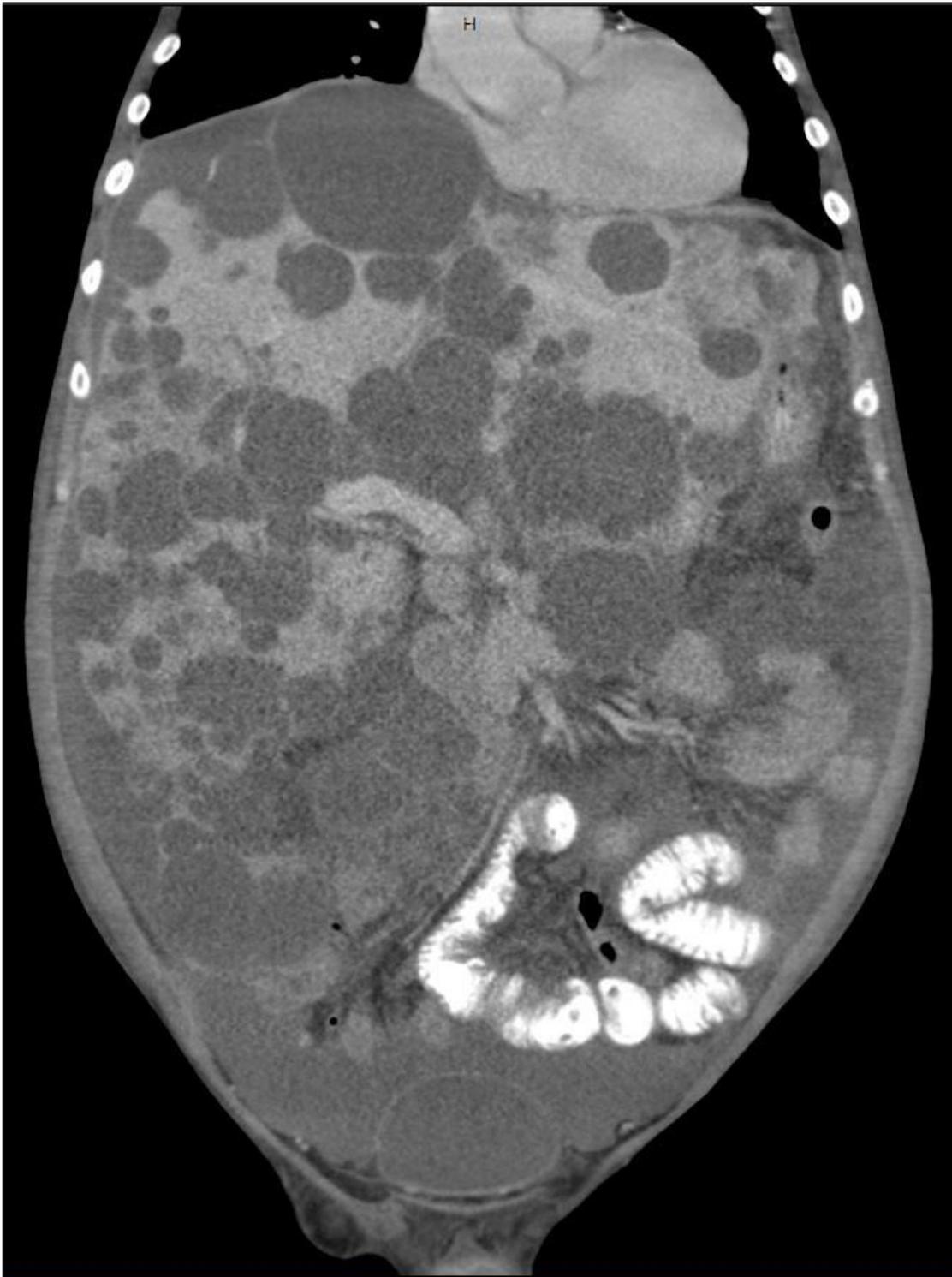


Figure 2

Coronal presentation of the portal vein in a polycystic liver. The kidneys are also polycystically degenerated as seen in the CT-scan (CT Revolution 256 slice, GE Healthcare, USA) before transplantation.



Figure 3

Coronal presentation of the portal vein in a polycystic liver. The kidneys are also polycystically degenerated as seen in the CT-scan (CT Revolution 256 slice, GE Healthcare, USA) before transplantation.