ISSN 2307-8960 (online)

World Journal of *Clinical Cases*

World J Clin Cases 2023 February 16; 11(5): 979-1223

Published by Baishideng Publishing Group Inc

World Journal of Clinical Cases

Contents

Thrice Monthly Volume 11 Number 5 February 16, 2023

MINIREVIEWS

979 Non-clostridium difficile induced pseudomembranous colitis

Jagirdhar GSK, Surani S

989 Pleural effusion in critically ill patients and intensive care setting

Bediwy AS, Al-Biltagi M, Saeed NK, Bediwy HA, Elbeltagi R

ORIGINAL ARTICLE

Retrospective Study

1000 Investigation of litigation in trauma orthopaedic surgery

Salimi M, Heidari MB, Ravandi Z, Mosalamiaghili S, Mirghaderi P, Jafari Kafiabadi M, Biglari F, Salimi A, Sabaghzadeh Irani A, Khabiri SS

1009 Type 2 diabetes mellitus characteristics affect hepatocellular carcinoma development in chronic hepatitis B patients with cirrhosis

Li MY, Li TT, Li KJ, Zhou C

1019 Relationship between glycemic variability and cognitive function in lacune patients with type 2 diabetes

Meng QZ, Wang Y, Li B, Xi Z, Wang M, Xiu JQ, Yang XP

1031 COVID-19-related cardiomyopathy: Can dual-energy computed tomography be a diagnostic tool?

Aydin F, Kantarci M, Aydın S, Karavaş E, Ceyhun G, Ogul H, Şahin ÇE, Eren S

Observational Study

1040 Multiple regression analysis of risk factors related to radiation pneumonitis

Shi LL, Yang JH, Yao HF

1049 Right hemicolectomy combined with duodenum-jejunum Roux-en-Y anastomosis for hepatic colon carcinoma invading the duodenum: A single-center case series

Liu PG, Feng PF, Chen XF

1058 Analysis of the value and safety of thyroid-stimulating hormone in the clinical efficacy of patients with thyroid cancer

Liang JJ, Feng WJ, Li R, Xu RT, Liang YL

CASE REPORT

1068 Effect of liver transplantation with primary hyperoxaluria type 1: Five case reports and review of literature

Wang XY, Zeng ZG, Zhu ZJ, Wei L, Qu W, Liu Y, Tan YL, Wang J, Zhang HM, Shi W, Sun LY

1077 Diagnosis of an intermediate case of maple syrup urine disease: A case report

Lin YT, Cai YN, Ting TH, Liu L, Zeng CH, Su L, Peng MZ, Li XZ

World Journal of Clinical Cases

Thrice Monthly Volume 11 Number 5 February 16, 2023

Contents

1086 Angioimmunoblastic T-cell lymphoma induced hemophagocytic lymphohistiocytosis and disseminated intravascular coagulopathy: A case report

Jiang M, Wan JH, Tu Y, Shen Y, Kong FC, Zhang ZL

1094 Giant myxofibrosarcoma of the esophagus treated by endoscopic submucosal dissection: A case report

Wang XS, Zhao CG, Wang HM, Wang XY

1099 Novel gene mutation in maturity-onset diabetes of the young: A case report

Zhang N, Zhao H, Li C, Zhang FZ

1106 Orthodontic-surgical treatment for severe skeletal class II malocclusion with vertical maxillary excess and four premolars extraction: A case report

Zhou YW, Wang YY, He ZF, Lu MX, Li GF, Li H

1115 Envafolimab combined with chemotherapy in the treatment of combined small cell lung cancer: A case report

Liu MH, Li YX, Liu Z

1122 Thyrotoxicosis in patients with a history of Graves' disease after SARS-CoV-2 vaccination (adenovirus vector vaccine): Two case reports

Yan BC, Luo RR

1129 Administration of modified Gegen Qinlian decoction for hemorrhagic chronic radiation proctitis: A case report and review of literature

Liu SY, Hu LL, Wang SJ, Liao ZL

1137 Surgical resection of a giant thymolipoma causing respiratory failure: A case report

Gong LH, Wang WX, Zhou Y, Yang DS, Zhang BH, Wu J

1144 Successful treatment of granulomatosis with polyangiitis using tocilizumab combined with glucocorticoids: A case report

Tang PF, Xu LC, Hong WT, Shi HY

1152 Langerhans cell histiocytosis misdiagnosed as thyroid malignancy: A case report

Shi JJ, Peng Y, Zhang Y, Zhou L, Pan G

1158 Combined treatment of refractory benign stricture after esophageal endoscopic mucosal dissection: A case report

Pu WF, Zhang T, Du ZH

1165 Bladder preservation in complicated invasive urothelial carcinoma following treatment with cisplatin/gemcitabine plus tislelizumab: A case report

Yang R, Chen JX, Luo SH, Chen TT, Chen LW, Huang B

1175 *Nocardia cyriacigeorgica* infection in a patient with repeated fever and $CD4^+$ T cell deficiency: A case report

Hong X, Ji YQ, Chen MY, Gou XY, Ge YM

World Journal of Clinical Cases

Thrice Monthly Volume 11 Number 5 February 16, 2023

Contents

1182 Closed loop ileus caused by a defect in the broad ligament: A case report

Zucal I, Nebiker CA

1188 Early postsurgical lethal outcome due to splenic littoral cell angioma: A case report

Jia F, Lin H, Li YL, Zhang JL, Tang L, Lu PT, Wang YQ, Cui YF, Yang XH, Lu ZY

1198 Combinations of nerve blocks in surgery for post COVID-19 pulmonary sequelae patient: A case report and review of literature

Jin Y, Lee S, Kim D, Hur J, Eom W

1206 Incidental right atrial mass in a patient with secondary pancreatic cancer: A case report and review of literature

Fioretti AM, Leopizzi T, La Forgia D, Scicchitano P, Oreste D, Fanizzi A, Massafra R, Oliva S

1217 Difficult airway due to cervical haemorrhage caused by spontaneous rupture of a parathyroid adenoma: A case report

Han YZ, Zhou Y, Peng Y, Zeng J, Zhao YQ, Gao XR, Zeng H, Guo XY, Li ZQ

World Journal of Clinical Cases

Thrice Monthly Volume 11 Number 5 February 16, 2023

Contents

ABOUT COVER

Editorial Board Member of *World Journal of Clinical Cases*, Tian-Biao Zhou, MD, PhD, Chief Doctor, Professor, Department of Nephrology, Second Affiliated Hospital, Shantou University Medical College, Shantou 515041, Guangdong Province, China. zhoutb@aliyun.com

AIMS AND SCOPE

The primary aim of *World Journal of Clinical Cases* (*WJCC*, *World J Clin Cases*) is to provide scholars and readers from various fields of clinical medicine with a platform to publish high-quality clinical research articles and communicate their research findings online.

WJCC mainly publishes articles reporting research results and findings obtained in the field of clinical medicine and covering a wide range of topics, including case control studies, retrospective cohort studies, retrospective studies, clinical trials studies, observational studies, prospective studies, randomized controlled trials, randomized clinical trials, systematic reviews, meta-analysis, and case reports.

INDEXING/ABSTRACTING

The *WJCC* is now abstracted and indexed in Science Citation Index Expanded (SCIE, also known as SciSearch®), Journal Citation Reports/Science Edition, Current Contents®/Clinical Medicine, PubMed, PubMed Central, Scopus, Reference Citation Analysis, China National Knowledge Infrastructure, China Science and Technology Journal Database, and Superstar Journals Database. The 2022 Edition of Journal Citation Reports® cites the 2021 impact factor (IF) for *WJCC* as 1.534; IF without journal self cites: 1.491; 5-year IF: 1.599; Journal Citation Indicator: 0.28; Ranking: 135 among 172 journals in medicine, general and internal; and Quartile category: Q4. The *WJCC*'s CiteScore for 2021 is 1.2 and Scopus CiteScore rank 2021: General Medicine is 443/826.

RESPONSIBLE EDITORS FOR THIS ISSUE

NAME OF JOURNAL
World Journal of Clinical Cases

ISSN
ISSN 2307-8960 (online)

LAUNCH DATE
April 16, 2013

FREQUENCY
Thrice Monthly

PUBLICATION DATE
February 16, 2023

World Journal of Clinical Cases

Submit a Manuscript: https://www.f6publishing.com

World J Clin Cases 2023 February 16; 11(5): 1068-1076

DOI: 10.12998/wjcc.v11.i5.1068

ISSN 2307-8960 (online)

CASE REPORT

Effect of liver transplantation with primary hyperoxaluria type 1: Five case reports and review of literature

Xin-Yue Wang, Zhi-Gui Zeng, Zhi-Jun Zhu, Lin Wei, Wei Qu, Ying Liu, Yu-Le Tan, Jun Wang, Hai-Ming Zhang, Wen Shi, Li-Ying Sun

Specialty type: Medicine, research and experimental

Provenance and peer review: Unsolicited article; Externally peer reviewed

Peer-review model: Single blind

Peer-review report's scientific quality classification

Grade A (Excellent): 0
Grade B (Very good): 0
Grade C (Good): C, C
Grade D (Fair): 0
Grade E (Poor): 0

P-Reviewer: Gupta R, India; Tuysuz U, Turkey

Received: October 9, 2022
Peer-review started: October 9, 2022
First decision: November 4, 2022
Revised: November 22, 2022
Accepted: January 9, 2023
Article in press: January 9, 2023
Published online: February 16, 2023

Xin-Yue Wang, Ying Liu, Li-Ying Sun, Department of Critical Liver Diseases, Liver Research Center, Beijing Friendship Hospital, Capital Medical University, Beijing 101100, China

Xin-Yue Wang, Zhi-Gui Zeng, Zhi-Jun Zhu, Lin Wei, Wei Qu, Ying Liu, Yu-Le Tan, Jun Wang, Hai-Ming Zhang, Li-Ying Sun, Liver Transplantation Center, Beijing Friendship Hospital, Capital Medical University, Beijing 101100, China

Xin-Yue Wang, Zhi-Gui Zeng, Zhi-Jun Zhu, Lin Wei, Wei Qu, Ying Liu, Yu-Le Tan, Jun Wang, Hai-Ming Zhang, Li-Ying Sun, Clinical Center for Pediatric Liver Transplantation, Capital Medical University, Beijing 101100, China

Xin-Yue Wang, Zhi-Gui Zeng, Zhi-Jun Zhu, Lin Wei, Wei Qu, Ying Liu, Yu-Le Tan, Jun Wang, Hai-Ming Zhang, Li-Ying Sun, National Clinical Research Center for Digestive Diseases, Beijing Friendship Hospital, Capital Medical University, Beijing 101100, China

Wen Shi, Beijing Clinical Research Institute, Beijing Friendship Hospital, Capital Medical University, Beijing 100050, China

Corresponding author: Li-Ying Sun, MD, Professor, Department of Critical Liver Diseases, Liver Research Center, Beijing Friendship Hospital, Capital Medical University, No. 101 Luyuandong Road, Tongzhou District, Beijing 101100, China. sunxlx@outlook.com

Abstract

BACKGROUND

Primary hyperoxaluria type 1 (PH1) is a rare autosomal recessive disease stemming from a deficiency in liver-specific alanine-glyoxylate aminotransferase, resulting in increased endogenous oxalate deposition and end-stage renal disease. Organ transplantation is the only effective treatment. However, its approach and timing remain controversial.

CASE SUMMARY

We retrospectively analyzed 5 patients diagnosed with PH1 from the Liver Transplant Center of the Beijing Friendship Hospital from March 2017 to December 2020. Our cohort included 4 males and 1 female. The median age at onset was 4.0 years (range: 1.0-5.0), age at diagnosis was 12.2 years (range: 6.7-23.5), age at liver transplantation (LT) was 12.2 years (range: 7.0-25.1), and the follow-up time was 26.3 mo (range: 12.8-40.1). All patients had delayed diagnosis, and 3 patients had progressed to end-stage renal disease by the time they were

diagnosed. Two patients received preemptive LT; their estimated glomerular filtration rate was maintained at > 120 mL/min/1.73 m^2, indicating a better prognosis. Three patients received sequential liver and kidney transplantation. After transplantation, serum and urinary oxalate decreased, and liver function recovered. At the last follow-up, the estimated glomerular filtration rates of the latter 3 patients were 179, 52 and 21 mL/min/1.73 m^2.

CONCLUSION
Different transplantation strategies should be adopted for patients based on their renal function stage. Preemptive-LT offers a good therapeutic approach for PH1.

Key Words: Primary hyperoxaluria type 1; Liver transplantation; Combined liver and kidney transplantation; Sequential liver and kidney transplantation; Renal calculi; End-stage renal disease; Case reports

Core Tip: Primary hyperoxaluria type 1 (PH1) is a rare autosomal recessive disease, resulting in increased endogenous oxalate deposition and end-stage renal disease. Organ transplantation is the only effective treatment. However, the approach and timing are controversial. To investigate the effect and timing of liver transplantation (LT) for PH1, we retrospectively analyzed 5 patients. The conclusions were that LT can treat PH1, and different transplantation strategies should be adopted for patients with different renal function stages. Preemptive-LT is a more appropriate treatment.

Citation: Wang XY, Zeng ZG, Zhu ZJ, Wei L, Qu W, Liu Y, Tan YL, Wang J, Zhang HM, Shi W, Sun LY. Effect of liver transplantation with primary hyperoxaluria type 1: Five case reports and review of literature. *World J Clin Cases* 2023; 11(5): 1068-1076
URL: https://www.wjgnet.com/2307-8960/full/v11/i5/1068.htm
DOI: https://dx.doi.org/10.12998/wjcc.v11.i5.1068

INTRODUCTION

Primary hyperoxaluria (PH) is a rare autosomal recessive disease with three subtypes. A deficiency of liver-specific alanine-glyoxylate aminotransferase, resulting in increased endogenous oxalate deposition, results in hyperoxaluria[1,2]. Because of the rarity and high rate of misdiagnosis of this condition, most patients present with existing end-stage renal disease (ESRD). Based on existing literature, organ transplantation is the only effective treatment for PH[3]. Liver transplantation (LT), as one of the treatments for genetic metabolic diseases, can result in partial or full remission and prevent metabolic crises[4]. Since 2013, more than 200 cases of genetic metabolic diseases have been treated at our LT center. Some of these diseases included abnormal organic acid metabolism[5], urea cycle disorder and PH. The surgeries performed included living donor LT, post-mortem organ donation and cross-auxiliary LT[6,7]. The following is our experience in the Beijing Friendship Hospital liver transplantation center of 5 cases with PH1 seen from 2017 to 2020. In this report, we provided data on quantified oxalic acid (OA) concentrations in serum and urine, which have been rarely reported in the literature and discuss the timing and effects of transplantation.

CASE PRESENTATION

Chief complaints

Case 1: Bilateral renal calculi for 2 years and PH1 diagnosis for 7 mo.

Case 2: Bilateral renal calculi for 22 years, nausea and vomiting for 1 year and PH1 diagnosis for 7 mo.

Case 3: Bilateral renal calculi for 15 years and PH1 diagnosis for 2 mo.

Case 4: Bilateral renal calculi for 11 years.

Case 5: Bilateral renal calculi for 5 years and decreased urine output for 16 d.

History of present illness

Case 1: Two years prior, bilateral renal calculi were identified by physical examination. There were no specific symptoms, and no treatment was prescribed. One month prior, the patient visited our hospital for analysis of gout/calculus factor in urine, which suggested oxaluria; genetic testing suggested PH1.

Case 2: Bilateral renal calculi were identified on a physical examination 22 years ago. The patient was prescribed medication to assist the passage of the calculi, but this failed. No further diagnosis or treatment was provided. Over a year prior, she developed anorexia, nausea and vomiting without obvious cause, and the symptoms gradually worsened. Hemoglobin (HGB) was 85 g/L, blood urea nitrogen was 36.8 mmol/L, and creatinine (Cr) was 1262.4 μmol/L from tests in another hospital. A bilateral renal ultrasound showed bilateral renal atrophy and multiple diffuse multiple renal calculi bilaterally. PH1 was diagnosed by genetic testing. The patient was put on hemodialysis (HD).

Case 3: No obvious cause was identified for abdominal pain, nausea and vomiting over 15 years. There were no other symptoms. The local hospital conducted an abdominal ultrasound, which identified renal calculi. Laparotomy was performed in a local hospital. After that, two additional extracorporeal shock wave lithotripsy were performed because of repeated stone discovery. Nausea and vomiting recommenced before 6 mo had elapsed. Blood Cr was 1465.7 mmol/L, and blood urea nitrogen was 36 mmol/L and the patient received HD. Genetic testing suggested PH1.

Case 4: The patient was diagnosed with two renal calculi 11 years prior. There was no extraordinary discomfort, and no treatment was provided. Genetic testing 2 mo prior suggested PH1.

Case 5: The patient was diagnosed with bilateral renal calculi 5 years prior without symptoms or treatment. One month prior, the patient experienced pain in the right side of the abdomen, nausea and vomiting. A right ureteral calculi and right ureteral stone were observed. There was no obvious cause of the reduced urine output (< 500 mL/d); light blood was noted in the urine. Serum Cr was 1300.0 μmol/L, and an abdominal ultrasound showed bilateral renal calculi and chronic kidney damage. Genetic testing suggested PH1, and the patient received HD.

History of past illness

Case 1: Myocarditis.

Cases 2 and 3: Hypertension.

Cases 4 and 5: None noted.

Personal and family history

All cases have none abnormalities detected.

Physical examination

All cases have none abnormalities detected.

Laboratory examinations

All cases were diagnosed as PH by genetic testing.

Case 1: HGB was 139 g/L, serum Cr was 81.0 μmol/L, and estimated glomerular filtration rate (eGFR) was 55.1 mL/min/ m^2.

Case 2: HGB was 85 g/L, serum Cr was 766.0 μmol/L, and eGFR was 5.8 mL/min/m^2.

Case 3: HGB was 71 g/L, serum Cr was 1541.0 μmol/L, and eGFR was 5.7 mL/min/m^2.

Case 4: HGB was 127 g/L, serum Cr was 80.0 μmol/L, and eGFR was 71.0 mL/min/m^2.

Case 5: HGB was 64 g/L, serum Cr was 967.9 μmol/L, and eGFR was 6.4 mL/min/m^2.

Imaging examinations

Cases 1, 2 and 4: Urinary system ultrasonography identified multiple calculi bilaterally.

Case 3: Urinary system ultrasonography identified multiple calculi bilaterally and nephrocalcalgia.

Case 5: Urinary system ultrasonography identified multiple calculi bilaterally and right ureteral calculus.

FINAL DIAGNOSIS

All cases: PH1, multiple calculi in both kidneys.

Case 1: Chronic kidney damage [chronic kidney disease (CKD)3].

Case 2: Renal anemia, chronic kidney damage (CKD3).

Case 3: Nephrocalcalgia, renal anemia, chronic kidney damage (CKD5).

Case 4: Chronic kidney damage (CKD3).

Case 5: Right ureteral calculus, renal anemia, chronic kidney damage (CKD5).

TREATMENT

All cases: Symptomatic treatment.

Cases 1, 2, and 4: Preemptive (pre)-LT.

Cases 3 and 5: Lithotomy, sequential liver and kidney transplantation (SLKT), HD.

OUTCOME AND FOLLOW-UP

General information and clinical manifestations

Between January 2017 and December 2020, 5 patients were diagnosed with PH1 by identifying mutations of the *AGXT* gene. There were four males and one female. The median age of onset (the first time that the patient's relatives first noticed the relevant clinical manifestations) was at 4.0 years (range: 1.0-5.0), the median age at diagnosis was 11.9-years-old (range: 6.7-23.5), and the median age at LT was 12.2-years-old (range: 7.0-25.1). The median follow-up time was 26.3 mo (range: 12.8-40.1 mo). All cases had urolithiasis and/or nephrocalcinosis. ESRD occurred in 3 cases. All 5 patients had a delayed diagnosis.

Laboratory examination

Biochemical blood and urine indices are shown in Table 1. The average HGB was 97.2 ± 30.2 g/L, average serum K^+ was 4.8 ± 0.7 mmol/L, average Na^+ was 141.0 ± 1.1 mmol/L, and average Ca^{2+} was 2.4 ± 0.1 mmol/L. Average eGFR was 28.8 ± 28.4 mL/min/1.73 m^2, and urine OA (UOA)/Cr was 748.2 ± 153.6 μg/mg Cr.

Molecular biology

Nine heterozygous mutations were identified in the 5 cases (Figure 1). The mutations included c.28_c.29delCCinsA, c815_c.816insGA, c.32C>G, c.346G>A, c.33_34insC, c.824_825insAG, c.26dupC and c.473C>T, c.145A>C. None of the mutations were identified to be novel. Two patients produced the c.32C>G mutation, and two had the c.824_825insAG mutation.

Operative and postoperative data

Among the 5 cases, two received pre-LT and three received SLKT (LT and KT were performed as separate procedures). Four donors were mothers, and one was a donor after cardiac death. The median interval between LT and KT of cases 2, 3 and 5 was 4.3 mo (range: 2.3-9.8); cases 1 and 4 received an LT only. Serum OA (SOA) of cases 3 and 5 before LT were 134.0 μmol/L and 233.6 μmol/L, respectively, while those of cases 2 and 3 before KT were 452.0 μmol/L and 142.5 μmol/L, respectively. Cases 1, 2 and 4 did not have recorded values before the LT, and case 5 did not have a recorded value before the KT. The median intensive care unit stay after LT was 5 d (range: 3-18). Renal function improved postoperatively in all cases. Table 2 shows the details of the surgical procedure and relevant data. All cases were followed-up to December 16, 2020, showing 100% survival at a median follow-up time of 26.3 mo (range: 12.8-40.1).

The normal plasma oxalate (POx) concentration was 10 mmol/L, which should be controlled below 50 μmol/L during dialysis. The normal 24-h urinary oxalate concentration was less than 45 mg (< 0.5 mmol/24 h/1.73 m^2). The normal value of urinary oxalate/Cr was related to age: 5-12 year, 47-119 μg/mg (60–150 μmol/mmol) and > 12 year 1.6-63.7 μg/mg (2-80 μmol/mmol)[8]. We used the Oxalic Acid Colorimetric Assay Kit (MAK179; Sigma-Aldrich, St Louis, MO, United States) to test the SOA and the UOA in our cases. The longitudinal data are shown in Figures 2A and 2B. The monitoring of these indices was not uniform due to patient compliance, issues with transportation and other problems.

Table 1 Preoperative laboratory examination

	Case 1	Case 2	Case 3	Case 4	Case 5
eGFR in mL/min/1.73 m^2	55.1	5.8	5.7	71.0	6.4
HGB in g/L	139	85	71	127	64
K^+ in mmol/L	4.4	4.8	6.2	4.5	4.2
Na^+ in mmol/L	140.1	142.2	140.9	139.6	142.2
Ca^{2+} in mmol/L	2.4	2.2	2.4	2.5	2.4
Urine OA/Cr in μg/mg Cr	824.83	Anuria	596.01	609.21	962.65
Serum OA as LT/KT in μmol/L	-	-/452	134/142.5	-	233.6/-

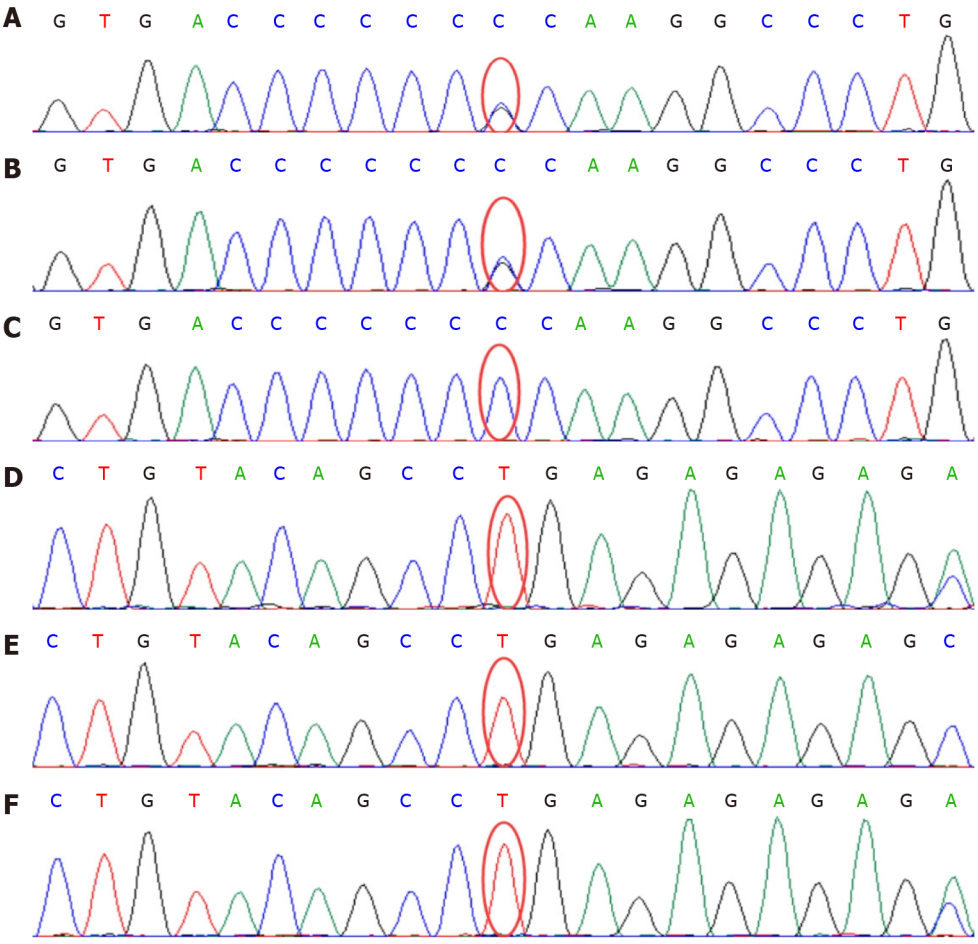
eGFR: Estimated glomerular filtration rate; HGB: Hemoglobin; KT: Kidney transplantation; LT: Liver transplantation; OA: Oxalic acid; OA/Cr: Oxalic acid/creatinine.

Table 2 Operative and postoperative clinical data

	Case 1	Case 2	Case 3	Case 4	Case 5
Tx	Pre-LT	SLKT	SLKT	Pre-LT	SLKT
LT donor	Mother	DCD	Mother	Mother	Mother
Liver segments	Left lobe graft	Left lobe graft	Right lobe graft	Left lobe graft	Left lateral section
Interval between LT and KT in mo	-	2.3	4.3	-	9.8
Serum OA level before Tx as LT/KT in μmol/L	-	-/452.0	134.0/142.5	-	233.6/-
Time in ICU as LT/KT in d	4	5/-	6/11	3	18/5
Dialysis early postoperative period as LT/KT	-	CVVHDF 5 d, HD 5.5 mo/HD 2.5 mo	HD 4.0 mo/-	-	HD/HD
Follow-up time in mo	35.9	40.1	26.3	13.1	12.8
Early postoperative complications	-	Biliary leakage	Fungal infections, hepatic artery occlusion, biliary leakage	-	Pneumonia, rotavirus diarrhea
Complications during the follow-up	CMV infection, mycoplasma infection	-	CMV infection	Hilar bile duct stricture, intrahepatic biliary ducts dilatation, obstructive jaundice	-
Liver outcome	Functional	Functional	Functional	Functional	Functional
Kidney outcome	-	Functional	Functional	-	Functional
Patient survival	Alive	Alive	Alive	Alive	Alive
Acute rejection	-	-	-	-	-
Current eGFR in mL/min/1.73m^2	127.4	20.5	52.0	157.0	178.7

-: Did not happen or the data was not recorded. CMV: Cytomegalovirus; CVVHDF: Continuous veno-venous hemodiafiltration; DCD: Donation after cardiac death; eGFR: Estimated glomerular filtration rate; HD: Hemodialysis; ICU: Intensive care unit; KT: Kidney transplantation; LT: Liver transplantation; OA: Oxalic acid; Pre-LT: Preemptive liver transplantation; SLKT: Estimated glomerular filtration rate; Tx: Transplantation.

As seen in Figure 2A, case 4 with pre-LT showed some improvement in SOA after surgery, whereas case 1 could not be evaluated due to incomplete data; all three SLKT cases (2, 3 and 5) showed postoperative improvements. Pre-LT case 4 showed improved in UOA postoperatively, whereas patient 1 could not be evaluated due to incomplete data. In SLKT patients, case 5 showed improvement postoperatively, while cases 2 and 3 had poor postoperative results (Figure 2B). Cases 1 and 4 with pre-LT and case 5 with SLKT showed a return to normal eGFR postoperatively; cases 2 and 3 showed



DOI: 10.12998/wjcc.v11.i5.1068

Figure 1 Case 4 *AGTX* gene sequencing peak. A: Patient, c.32c>G (heterozygous); B: Paternal origin variation, c.32c>G (heterozygous); C: Wild type, patient's mother; D: Patient, c.824_825insAG (heterozygous); E: Wild type, patient's father; F: Maternal origin variation, c.824_825insAG (heterozygous).

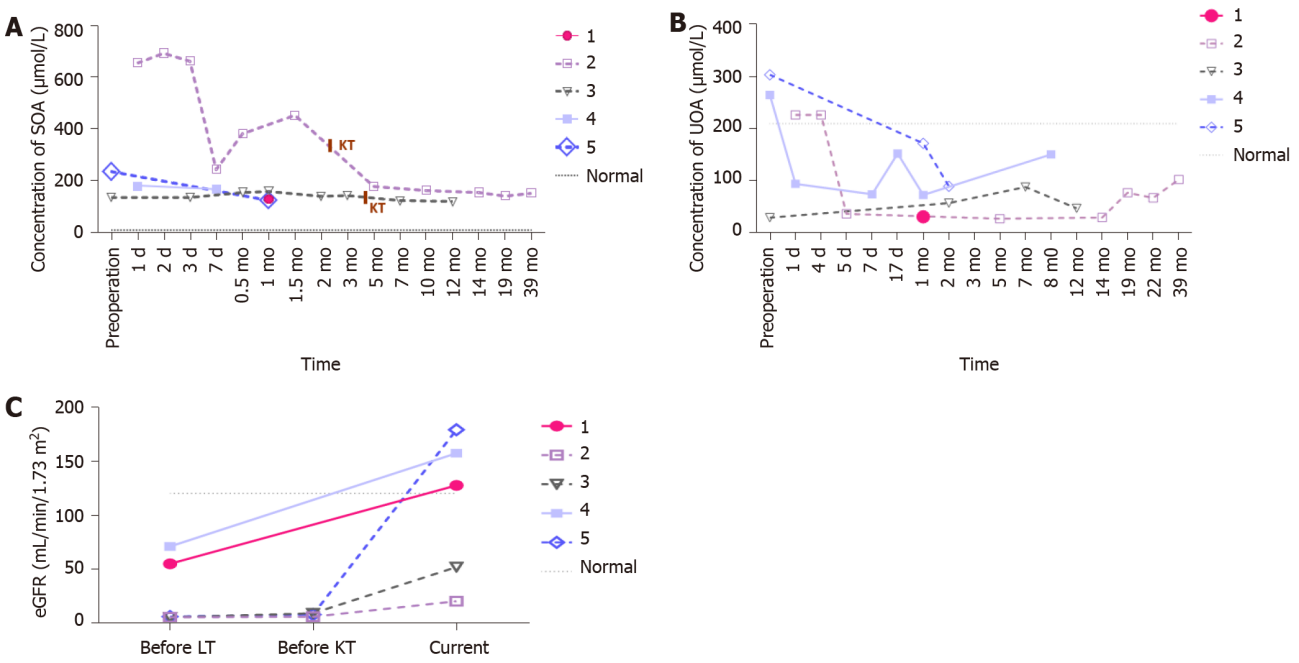
improvement. In terms of renal function, LT significantly improved renal function in cases 1 and 4 who underwent pre-LT. However, SLKT cases 2, 3 and 5 had little improvement of renal function at the LT stage, but improved renal function after KT; case 5 was the only patient to regain normal function.

DISCUSSION

PH is a rare autosomal recessive metabolic disease[9], stemming from a deficiency in liver-specific alanine-glyoxylate aminotransferase, which then results in endogenous oxalate production, urinary calculi and even renal dysfunction[10,11]. Of the three types of PH, PH1 is the most common, accounting for about 80% of all cases[8,12]. A study from Europe and North America reported an annual incidence rate of 0.1-0.2/1000000 and a prevalence rate of 0.8-2.9/1000000[8]. Due to the rarity of PH1, its diagnosis is often delayed or completely missed[13]. The median age of disease onset is 5.5-years-old or younger. Around 90% of these patients have calculi, and 20%-50% have developed ESRD by the time that a PH1 diagnosis is made[14].

Oxalate deposition is the critical saturation point of POx when GFR reduces below 60 mL/min/1.73 m^2. Systemic oxalosis occurs in many sites, including bone, heart, *etc*[11,15], and the immediate institution of HD is recommended. However, dialysis alone cannot overcome the continuous production of oxalate[16,17], leaving organ transplantation as the only effective therapy. However, transplantation strategies need to be tailored to individual patients[18-21]. Pre-LT should be performed in all patients in stage 3b of CKD (eGFR is 30-60 mL/min/1.73 m^2), and combined LT and KT should be performed when the patient is an infant or in CKD stage 4 (eGFR is 15-29 mL/min/1.73 m^2). SLKT is more suitable for patients with CKD stage 5 (eGFR < 15 mL/min/1.73 m^2, ESRD).

Combined LT and KT involves the implantation of a whole or partial transplanted liver and kidney from the same donor and during the same surgical procedure[22-24]. Continuous hyperhydration and HD are necessary[22,25-27]. However, dialysis cannot completely remove POx, and oxalation is often present in dialysis patients[19]. SLKT involves two stages: A single LT is performed followed by KT



DOI: 10.12998/wjcc.v11.i5.1068

Figure 2 Change of serum oxalic acid, urinary oxalic acid and estimated glomerular filtration rate. A: Serum oxalic acid concentration; B: Urinary oxalic acid concentration; C: Estimated glomerular filtration rate. eGFR: Estimated glomerular filtration rate; KT: Kidney transplantation; SOA: Serum oxalic acid; UOA: Urinary oxalic acid.

[23]. With this procedure, HD is maintained after LT until normal levels of oxalate are achieved, then the KT can proceed[17,21,28,29]. In our cases, deceased donors were used for case 2, while case 3 and 5 received their livers from a living donor and kidney from a deceased donor, with all three surviving at the last follow-up assessment. Following LT, the oxalate accumulates in the kidney, resulting in kidney disease. Therefore, intensive dialysis should be employed before KT to reduce POx below saturation levels[21]. As it is difficult to quantify the total oxalate load *in vivo*, there is no consensus on the optimal interval between LT and KT[19,21]. The normal ranges of plasma OA have been determined using different methods. Elgstoen[30] reported normal POA at 3-11 μmol/L (n = 67), while in their 3 PH patients these levels ranged from 50-170 μmol/L. In that study, OA was tested using the Oxalic Acid Colorimetric Assay Kit (MAK179; Sigma-Aldrich). To obtain control SOA levels in our study, we collected morning urine samples from 7 non-PH patients and found that in those individuals, urinary OA ranged from 69.7-208.0 μmol/L. Due to problems regarding patient compliance, family economic conditions, limited medical resources, *etc*, our 3 cases opted for conventional HD. The timing of the donor after cardiac death kidney donation was uncontrollable, and SOA did not decrease to normal before KT in the 3 cases. The postoperative recovery of SLKT in case 5 was superior to cases 2 and 3 based on renal function and UOA; this may be related to the shorter duration between ESRD development and transplantation (0.2 year in case 5 *vs* 2.9 year in case 2 and 0.6 year in case 3). In addition, case 5 received their organ from a living doner.

Pre-LT is an effective method for avoiding hepatic or renal failure[16,17,19]. There is no unified standard for conducting pre-LT when no obvious renal dysfunction is present[12]. Based on the recommendations of the Oxal Europe Expert Group[8], a pre-LT should be considered in patients with CKD stage 3b. Preoperative eGFRs in cases 1 and 4 were 55.1 and 71.0 mL/min/1.73 m^2, respectively, and both were staged as CKD stage 3. Their livers showed a good postoperative recovery. With regard to their calculi, case 1 remained relatively unchanged but case 4 showed an improvement; their latest eGFRs were 127.4 mL/min/1.73 m^2 and 157.0 mL/min/1.73 m^2, respectively, indicating a trend towards normal.

CONCLUSION

PH1 has a high rate of misdiagnosis, and high heterogeneity in terms of the clinical phenotype and genotype. Most patients develop ESRD by the time of their PH1 diagnosis. Each transplantation method has its advantages, which should be comprehensively considered as part of a tailored approach to treating the patient's condition and based on the abilities of the transplantation center and relevant economic factors. Both pre-LT and SLKT can be safely performed and provide encouraging results for patients with PH1. We believe that LT should be performed as early as possible to avoid further organ

damage and to maximize postoperative recovery.

FOOTNOTES

Author contributions: Wang XY and Sun LY designed the research study; Shi W and Wang XY performed the research; Wang XY analyzed the data and wrote the manuscript; Zeng ZG, Zhu ZJ, Wei L, Qu W, Liu Y, Tan YL, Wang J and Zhang HM performed the operations; and all authors have read and approved the final manuscript.

Informed consent statement: All study participants, or their legal guardian, provided informed written consent prior to study enrollment.

Conflict-of-interest statement: All authors report having no relevant conflicts of interest for this article.

CARE Checklist (2016) statement: The authors declare that the manuscript is checked with CARE Checklist (2016).

Country/Territory of origin: China

ORCID number: Zhi-Gui Zeng 0000-0003-1457-7495; Zhi-Jun Zhu 0000-0001-7031-2083; Lin Wei 0000-0002-0435-3829; Wei Qu 0000-0002-4484-5940; Ying Liu 0000-0001-9087-899X; Hai-Ming Zhang 0000-0003-4629-3913; Li-Ying Sun 0000-0003-1101-7994.

S-Editor: Wang JJ
L-Editor: Filipodia
P-Editor: Wang JJ

REFERENCES

1 **Leumann E**, Hoppe B. The primary hyperoxalurias. *J Am Soc Nephrol* 2001; **12**: 1986-1993 [PMID: 11518794 DOI: 10.1681/ASN.V1291986]
2 **Hoppe B**, Beck B, Gatter N, von Unruh G, Tischer A, Hesse A, Laube N, Kaul P, Sidhu H. Oxalobacter formigenes: a potential tool for the treatment of primary hyperoxaluria type 1. *Kidney Int* 2006; **70**: 1305-1311 [PMID: 16850020 DOI: 10.1038/sj.ki.5001707]
3 **Nolkemper D**, Kemper MJ, Burdelski M, Vaismann I, Rogiers X, Broelsch CE, Ganschow R, Müller-Wiefel DE. Long-term results of pre-emptive liver transplantation in primary hyperoxaluria type 1. *Pediatr Transplant* 2000; **4**: 177-181 [PMID: 10933316 DOI: 10.1034/j.1399-3046.2000.00107.x]
4 **Fagiuoli S**, Daina E, D'Antiga L, Colledan M, Remuzzi G. Monogenic diseases that can be cured by liver transplantation. *J Hepatol* 2013; **59**: 595-612 [PMID: 23578885 DOI: 10.1016/j.jhep.2013.04.004]
5 **Jiang YZ**, Sun LY, Zhu ZJ, Wei L, Qu W, Zeng ZG, Liu Y, Tan YL, He EH, Xu RF, Zhang L, Wang J, Chen XJ. Perioperative characteristics and management of liver transplantation for isolated methylmalonic acidemia-the largest experience in China. *Hepatobiliary Surg Nutr* 2019; **8**: 470-479 [PMID: 31673536 DOI: 10.21037/hbsn.2019.03.04]
6 **Qu W**, Wei L, Zhu ZJ, Sun LY, Liu Y, Zeng ZG. Considerations for Use of Domino Cross-auxiliary Liver Transplantation in Metabolic Liver Diseases: A Review of Case Studies. *Transplantation* 2019; **103**: 1916-1920 [PMID: 30801517 DOI: 10.1097/TP.0000000000002602]
7 **Zhu ZJ**, Wei L, Qu W, Sun LY, Liu Y, Zeng ZG, Zhang L, He EH, Zhang HM, Jia JD, Zhang ZT. First case of cross-auxiliary double domino donor liver transplantation. *World J Gastroenterol* 2017; **23**: 7939-7944 [PMID: 29209135 DOI: 10.3748/wjg.v23.i44.7939]
8 **Cochat P**, Rumsby G. Primary hyperoxaluria. *N Engl J Med* 2013; **369**: 649-658 [PMID: 23944302 DOI: 10.1056/NEJMra1301564]
9 **Salido E**, Pey AL, Rodriguez R, Lorenzo V. Primary hyperoxalurias: disorders of glyoxylate detoxification. *Biochim Biophys Acta* 2012; **1822**: 1453-1464 [PMID: 22446032 DOI: 10.1016/j.bbadis.2012.03.004]
10 **Cochat P**, Liutkus A, Fargue S, Basmaison O, Ranchin B, Rolland MO. Primary hyperoxaluria type 1: still challenging! *Pediatr Nephrol* 2006; **21**: 1075-1081 [PMID: 16810517 DOI: 10.1007/s00467-006-0124-4]
11 **Malde DJ**, Pararajasingam R, Tavakoli A, Campbell B, Riad H, Parrot N, Prasad KR, Augustine T. Transplantation in adults with primary hyperoxaluria: single unit experience and treatment algorithm. *Ann Transplant* 2011; **16**: 111-117 [PMID: 22210430 DOI: 10.12659/AOT.882227]
12 **Cochat P**, Groothoff J. Primary hyperoxaluria type 1: practical and ethical issues. *Pediatr Nephrol* 2013; **28**: 2273-2281 [PMID: 23494551 DOI: 10.1007/s00467-013-2444-5]
13 **Hoppe B**, Langman CB. A United States survey on diagnosis, treatment, and outcome of primary hyperoxaluria. *Pediatr Nephrol* 2003; **18**: 986-991 [PMID: 12920626 DOI: 10.1007/s00467-003-1234-x]

14 **Harambat J**, van Stralen KJ, Espinosa L, Groothoff JW, Hulton SA, Cerkauskiene R, Schaefer F, Verrina E, Jager KJ, Cochat P; European Society for Pediatric Nephrology/European Renal Association-European Dialysis and Transplant Association (ESPN/ERA-EDTA) Registry. Characteristics and outcomes of children with primary oxalosis requiring renal replacement therapy. *Clin J Am Soc Nephrol* 2012; **7**: 458-465 [PMID: 22223608 DOI: 10.2215/CJN.07430711]

15 **Soliman NA**, Nabhan MM, Abdelrahman SM, Abdelaziz H, Helmy R, Ghanim K, Bazaraa HM, Badr AM, Tolba OA, Kotb MA, Eweeda KM, Fayez A. Clinical spectrum of primary hyperoxaluria type 1: Experience of a tertiary center. *Nephrol Ther* 2017; **13**: 176-182 [PMID: 28161266 DOI: 10.1016/j.nephro.2016.08.002]

16 **Cochat P**, Hulton SA, Acquaviva C, Danpure CJ, Daudon M, De Marchi M, Fargue S, Groothoff J, Harambat J, Hoppe B, Jamieson NV, Kemper MJ, Mandrile G, Marangella M, Picca S, Rumsby G, Salido E, Straub M, van Woerden CS; OxalEurope. Primary hyperoxaluria Type 1: indications for screening and guidance for diagnosis and treatment. *Nephrol Dial Transplant* 2012; **27**: 1729-1736 [PMID: 22547750 DOI: 10.1093/ndt/gfs078]

17 **Harambat J**, Fargue S, Bacchetta J, Acquaviva C, Cochat P. Primary hyperoxaluria. *Int J Nephrol* 2011; **2011**: 864580 [PMID: 21748001 DOI: 10.4061/2011/864580]

18 **Beck BB**, Hoyer-Kuhn H, Göbel H, Habbig S, Hoppe B. Hyperoxaluria and systemic oxalosis: an update on current therapy and future directions. *Expert Opin Investig Drugs* 2013; **22**: 117-129 [PMID: 23167815 DOI: 10.1517/13543784.2013.741587]

19 **Alkhunaizi AM**, Al-Sannaa NA, Raslan WF. Hyperoxaluria and rapid development of renal failure following a combined liver and kidney transplantation: emphasis on sequential transplantation. *JIMD Rep* 2012; **3**: 91-95 [PMID: 23430879 DOI: 10.1007/8904_2011_67]

20 **Naderi G**, Tabassomi F, Latif A, Ganji M. The first experience of sequential liver-kidney transplantation for the treatment of primary hyperoxaluria type-1 in Iran as a developing country. *Saudi J Kidney Dis Transpl* 2016; **27**: 791-794 [PMID: 27424700 DOI: 10.4103/1319-2442.185262]

21 **Ozer A**, Aktas H, Bulum B, Emiroglu R. The experience of combined and sequential liver and kidney transplantation from a single living donor in patients with primary hyperoxaluria type 1. *Pediatr Transplant* 2019; **23**: e13406 [PMID: 30932299 DOI: 10.1111/petr.13406]

22 **Duclaux-Loras R**, Bacchetta J, Berthiller J, Rivet C, Demède D, Javouhey E, Dubois R, Dijoud F, Lachaux A, Badet L, Boillot O, Cochat P. Pediatric combined liver-kidney transplantation: a single-center experience of 18 cases. *Pediatr Nephrol* 2016; **31**: 1517-1529 [PMID: 27060059 DOI: 10.1007/s00467-016-3324-6]

23 **Grenda R**, Kaliciński P. Combined and sequential liver-kidney transplantation in children. *Pediatr Nephrol* 2018; **33**: 2227-2237 [PMID: 29322327 DOI: 10.1007/s00467-017-3880-4]

24 **Szymczak M**, Kaliciński P, Kowalewski G, Markiewicz-Kijewska M, Broniszczak D, Ismail H, Stefanowicz M, Kowalski A, Rubik J, Jankowska I, Piątosa B, Teisseyre J, Grenda R. Combined Liver-Kidney Transplantation in Children: Single-Center Experiences and Long-Term Results. *Transplant Proc* 2018; **50**: 2140-2144 [PMID: 30177126 DOI: 10.1016/j.transproceed.2018.04.061]

25 **Büscher R**, Büscher AK, Cetiner M, Treckmann JW, Paul A, Vester U, Hoyer PF. Combined liver and kidney transplantation and kidney after liver transplantation in children: Indication, postoperative outcome, and long-term results. *Pediatr Transplant* 2015; **19**: 858-865 [PMID: 26341656 DOI: 10.1111/petr.12595]

26 **Harps E**, Brinkert F, Ganschow R, Briem-Richter A, van Husen M, Schmidtke S, Herden U, Nashan B, Fischer L, Kemper MJ. Immediate postoperative intensive care treatment of pediatric combined liver-kidney transplantation: outcome and prognostic factors. *Transplantation* 2011; **91**: 1127-1131 [PMID: 21544033 DOI: 10.1097/TP.0b013e318216c1bb]

27 **Kemper MJ**. Concurrent or sequential liver and kidney transplantation in children with primary hyperoxaluria type 1? *Pediatr Transplant* 2005; **9**: 693-696 [PMID: 16269037 DOI: 10.1111/j.1399-3046.2005.00362.x]

28 **Kitajima K**, Ogawa Y, Miki K, Kai K, Sannomiya A, Iwadoh K, Murakami T, Koyama I, Nakajima I, Fuchinoue S. Longterm renal allograft survival after sequential liver-kidney transplantation from a single living donor. *Liver Transpl* 2017; **23**: 315-323 [PMID: 27862900 DOI: 10.1002/lt.24676]

29 **Malla I**, Lysy PA, Godefroid N, Smets F, Malaise J, Reding R, Sokal EM. Two-step transplantation for primary hyperoxaluria: cadaveric liver followed by living donor related kidney transplantation. *Pediatr Transplant* 2009; **13**: 782-784 [PMID: 19032423 DOI: 10.1111/j.1399-3046.2008.01049.x]

30 **Elgstoen KB**. Liquid chromatography-tandem mass spectrometry method for routine measurement of oxalic acid in human plasma. *J Chromatogr B Analyt Technol Biomed Life Sci* 2008; **873**: 31-36 [PMID: 18755640 DOI: 10.1016/j.jchromb.2008.07.002]

Published by **Baishideng Publishing Group Inc**
7041 Koll Center Parkway, Suite 160, Pleasanton, CA 94566, USA
Telephone: +1-925-3991568
E-mail: bpgoffice@wjgnet.com
Help Desk: https://www.f6publishing.com/helpdesk
https://www.wjgnet.com