



MedStar Georgetown  
Transplant Institute



# Transplant for MSUD

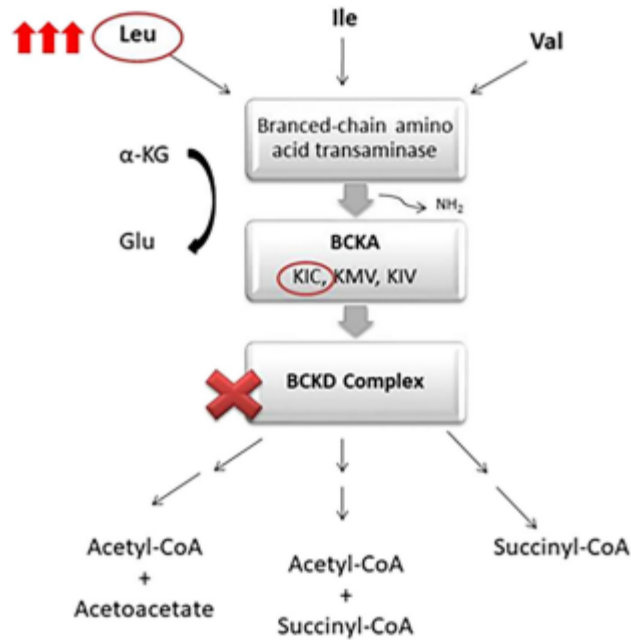
Juan Francisco Guerra, MD  
Transplant Surgeon

Medstar Georgetown Transplant Institute

# MSUD



# MSUD



Ketoacidosis  
Ataxia  
Coma  
Mental and psychomotor  
delay

FIGURE 1 Metabolic pathway of branched-chain amino acids leucine (Leu), isoleucine (Ile), and valine (Val), indicating the metabolic blockade that occurs in maple syrup urine disease (MSUD), located in the branched-chain  $\alpha$ -ketoacid dehydrogenase complex (BCKD complex).  $\alpha$ -Keto-isocaproic (KIC),  $\alpha$ -keto- $\beta$ -methyl valeric (KMV),  $\alpha$ -keto-isovaleric (KIV),  $\alpha$ -keto-glutarate ( $\alpha$ -KG), and glutamate (Glu). (adapted from Scriver et al., 2001 and Xu et al., 2020).

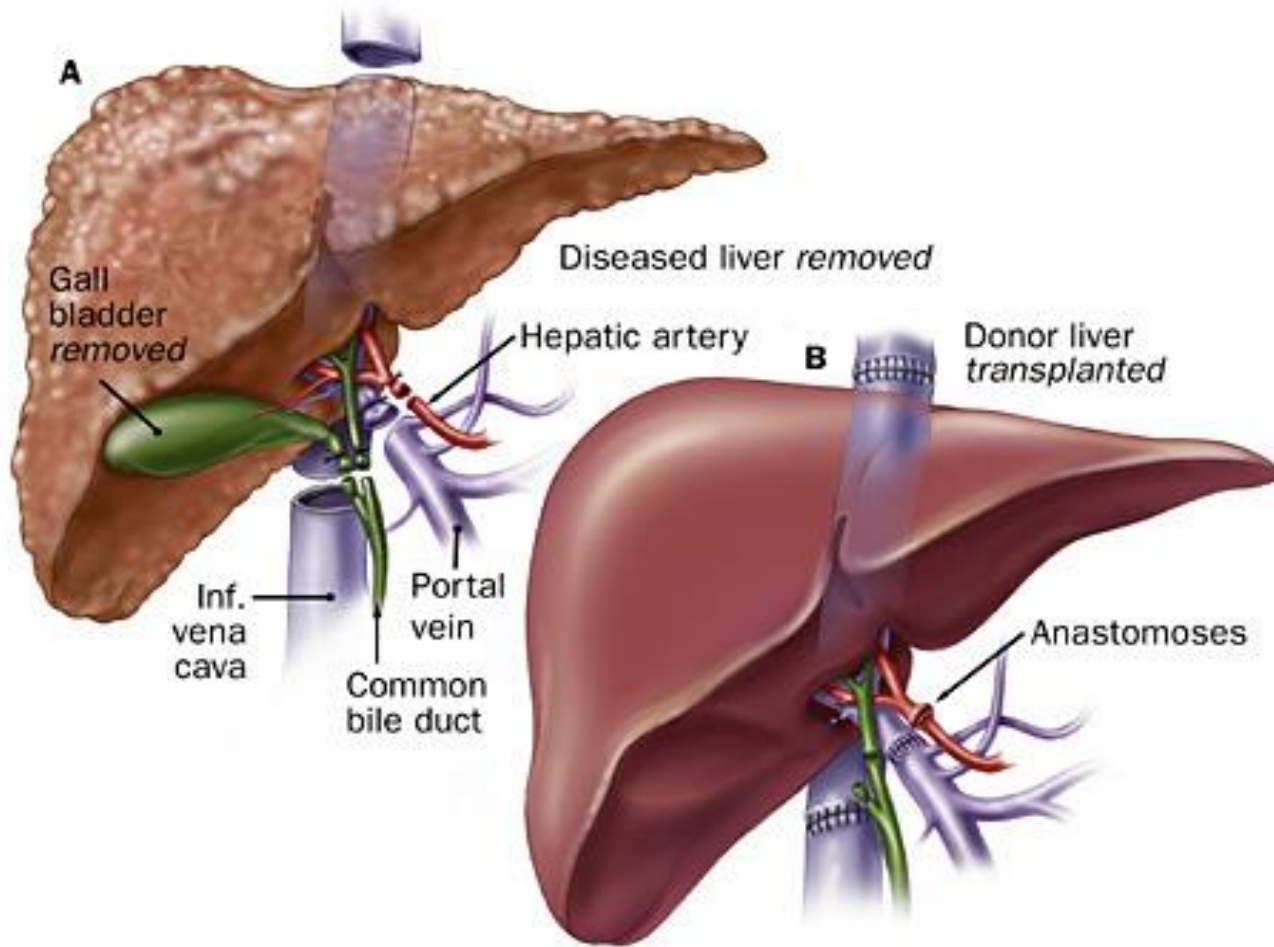
# Background

- Mechanisms not fully understood
- Early diagnosis and treatment
- Proper management of decomp  
  - “Standard treatment”



- Liver transplantation

# Surgical Considerations



# Background

- Different “metabolic” conditions
- First cases: NOT metabolic reasons



- Improved dietary restrictions
- Reduced levels of LEU, ISO and VAL
- More “manageable”

# Goal?

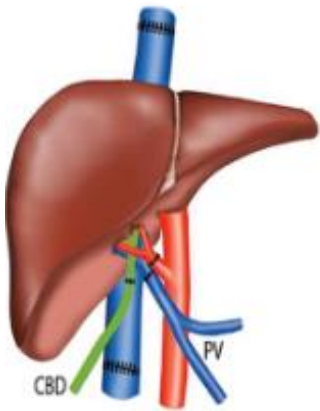
- Benefit for an individual patient?



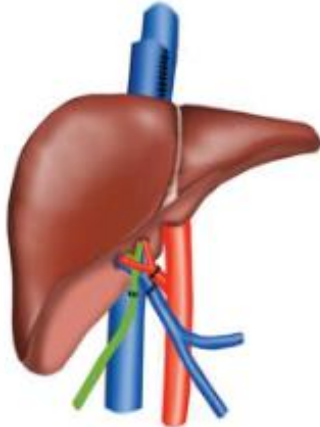
- More “manageable” disease v/s risk of Immunosuppression

# Surgical Considerations

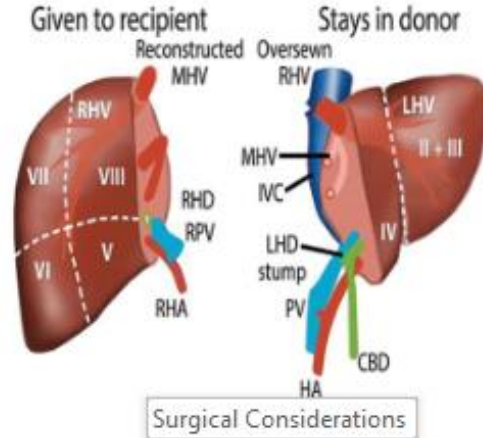
**A. Conventional technique**



**B. Piggyback technique**

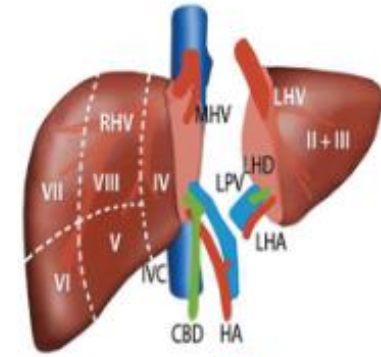


**C. Living donor right lobe liver transplantation**

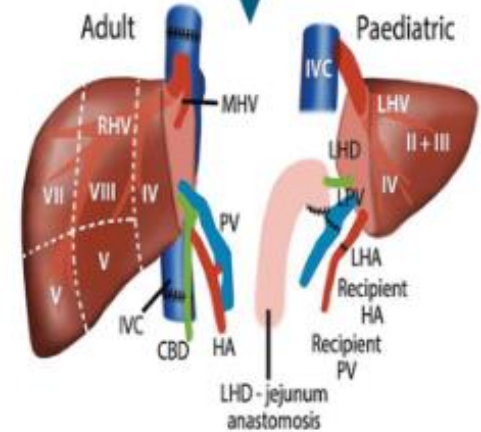
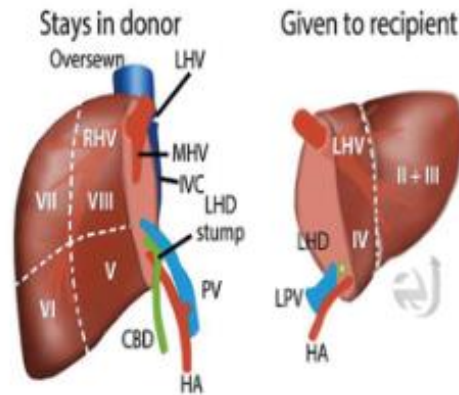


Surgical Considerations

**D. Split liver**



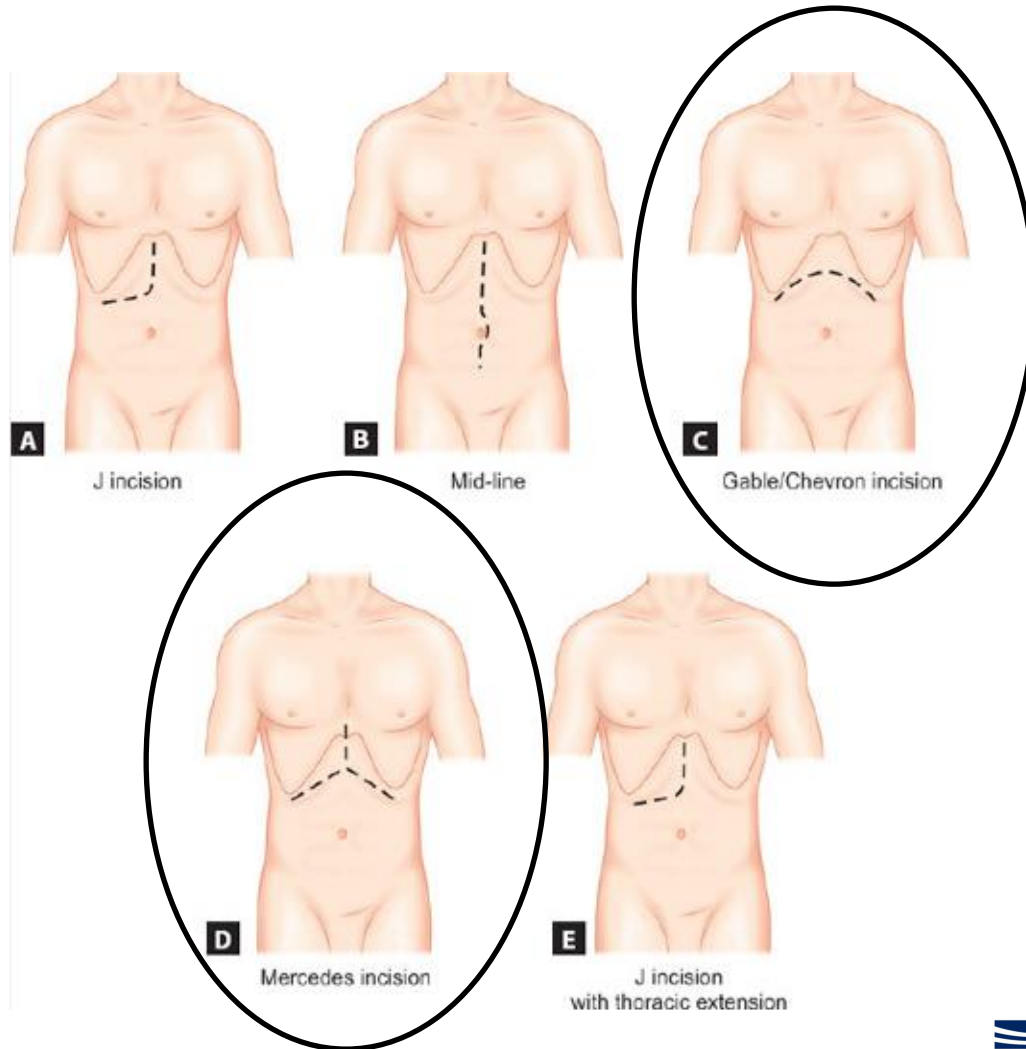
**E. Living donor left lobe liver transplantation**



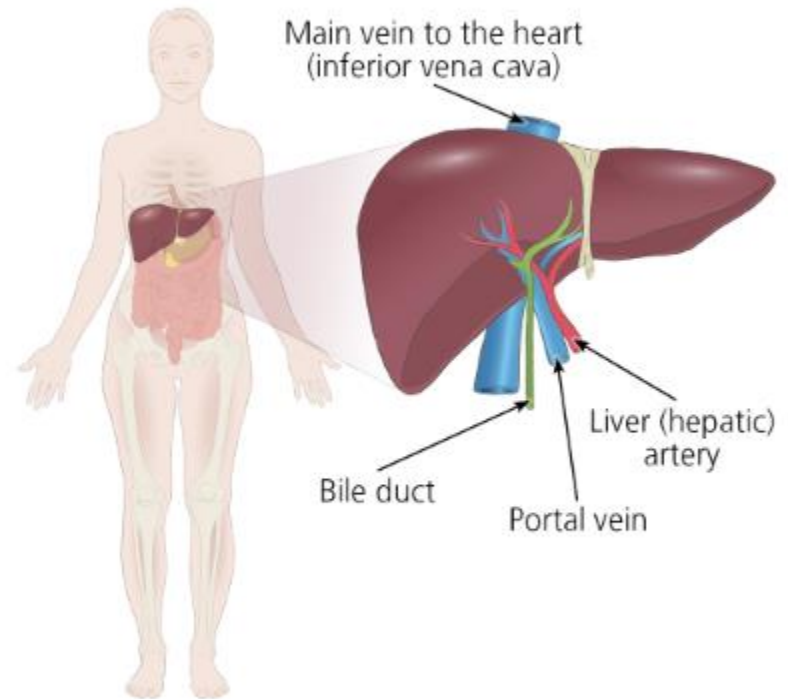
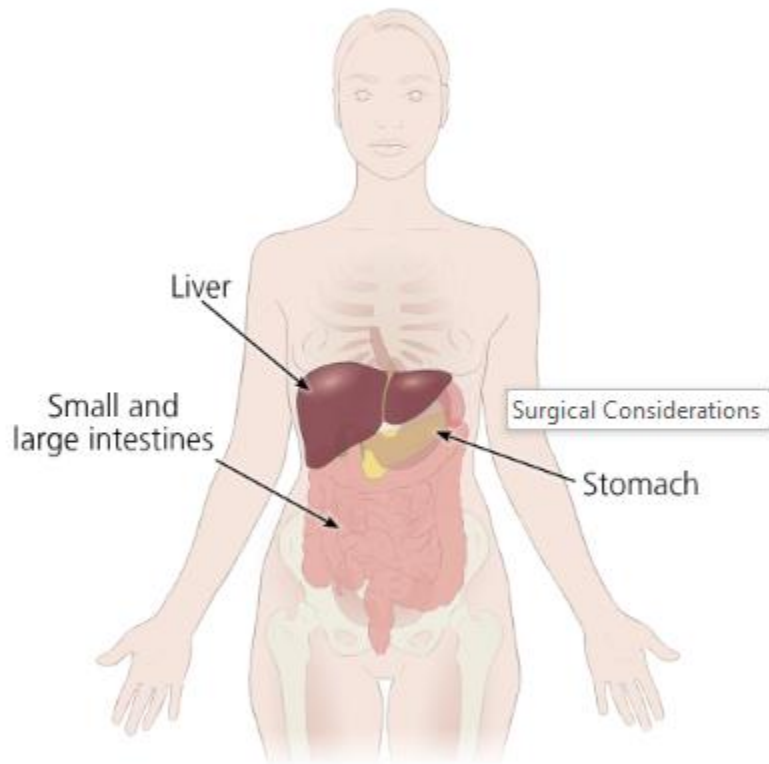
- IVC Inferior vena cava
- HA Hepatic artery
- PV Portal vein
- CBD Common bile duct
- LHA Left hepatic artery
- LPV Left portal vein
- LHD Left hepatic duct
- LHV Left hepatic vein
- RHA Right hepatic artery
- RPV Right portal vein
- RHD Right hepatic duct
- RHV Right hepatic vein
- MHV Middle hepatic vein



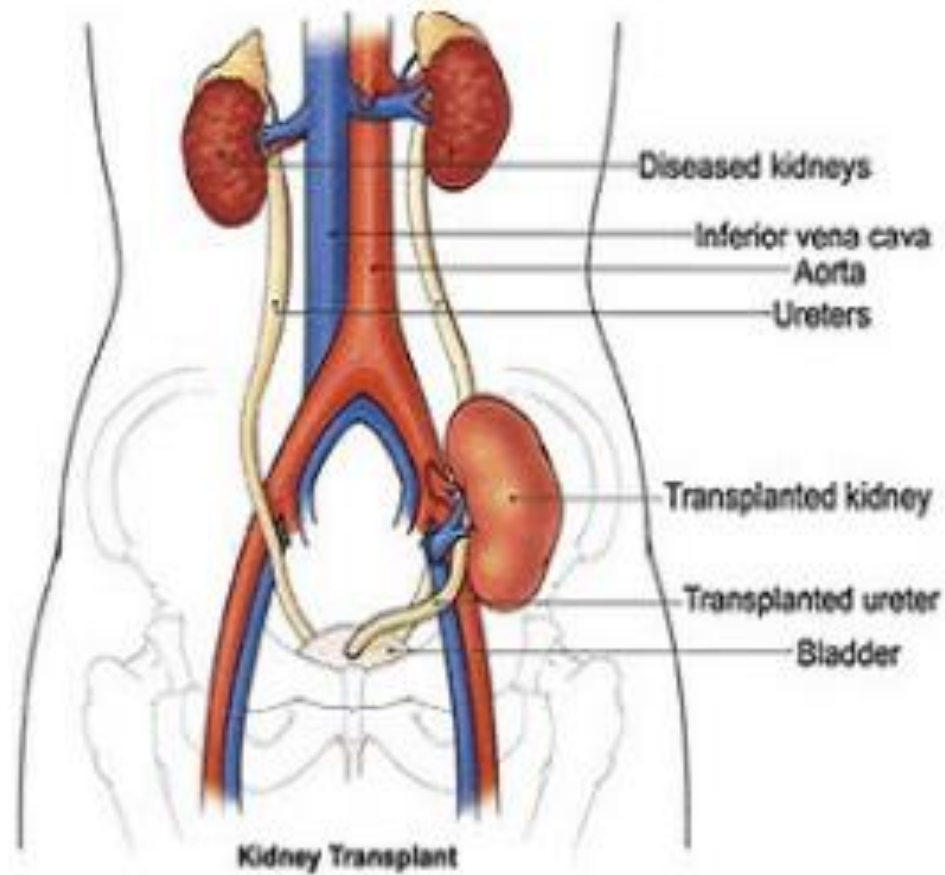
# Surgical Considerations



# Surgical Considerations



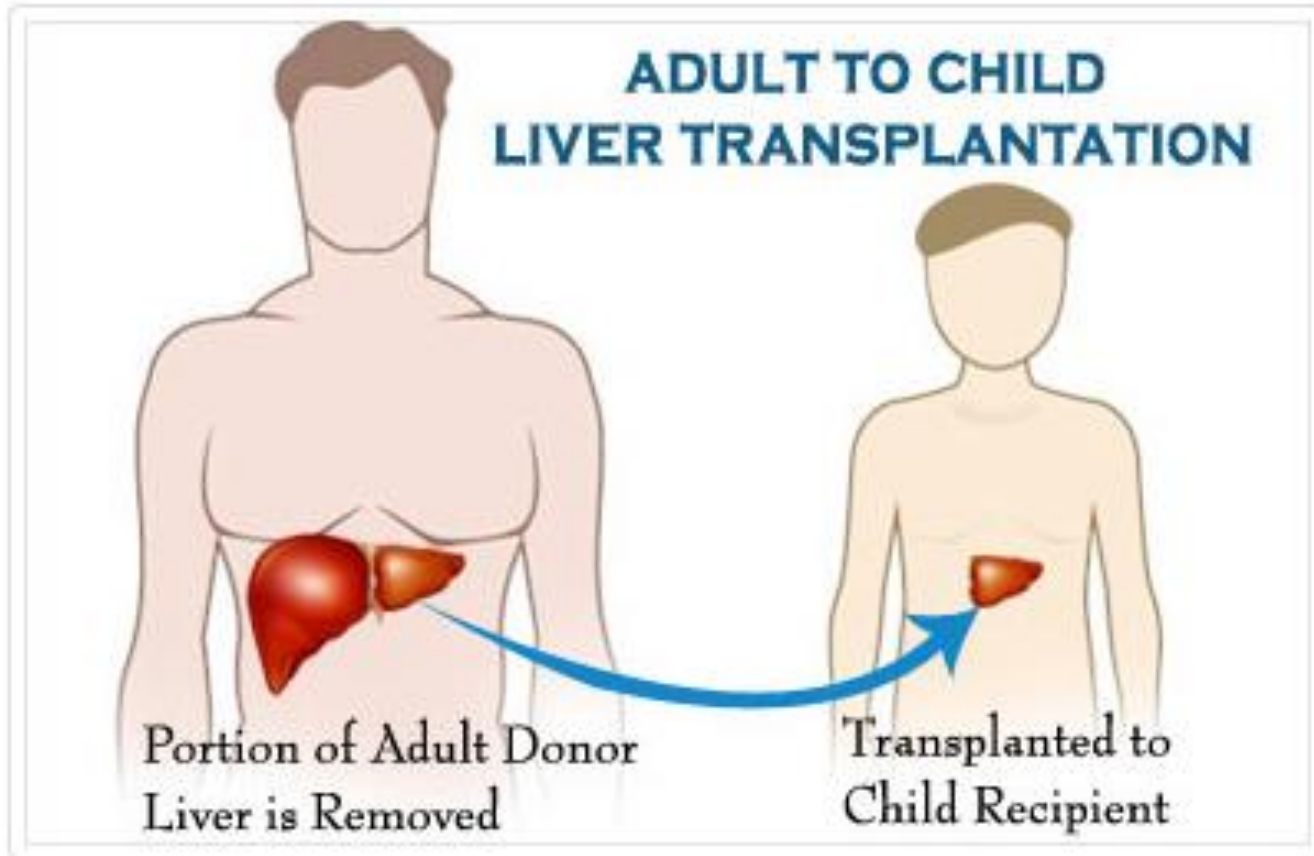
# Kidney Transplant



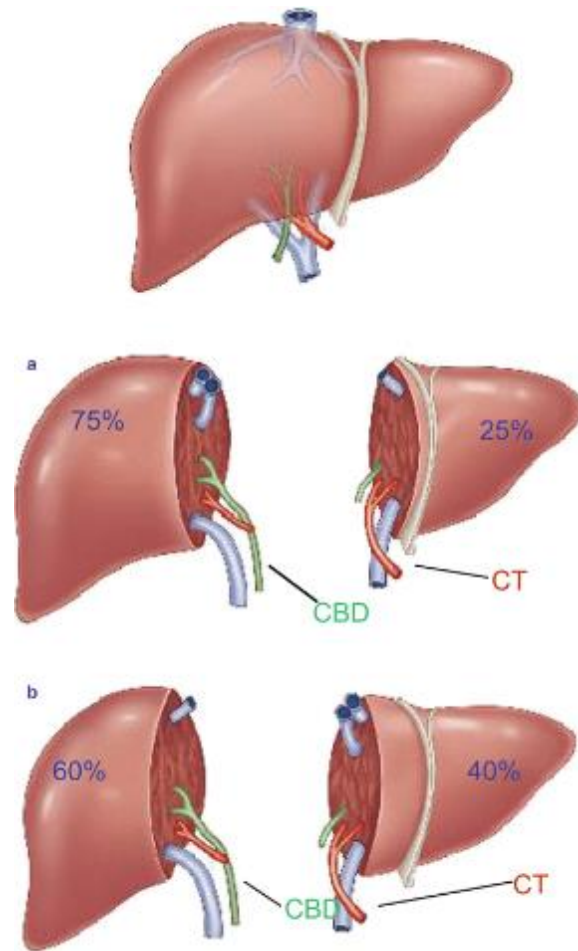
# Deceased donor



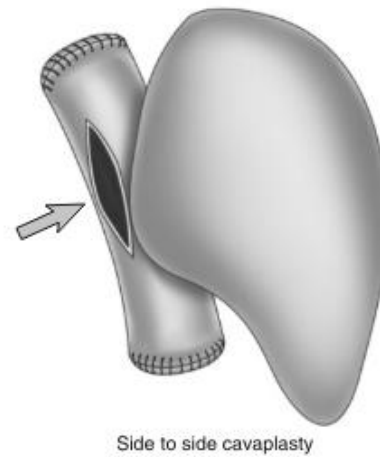
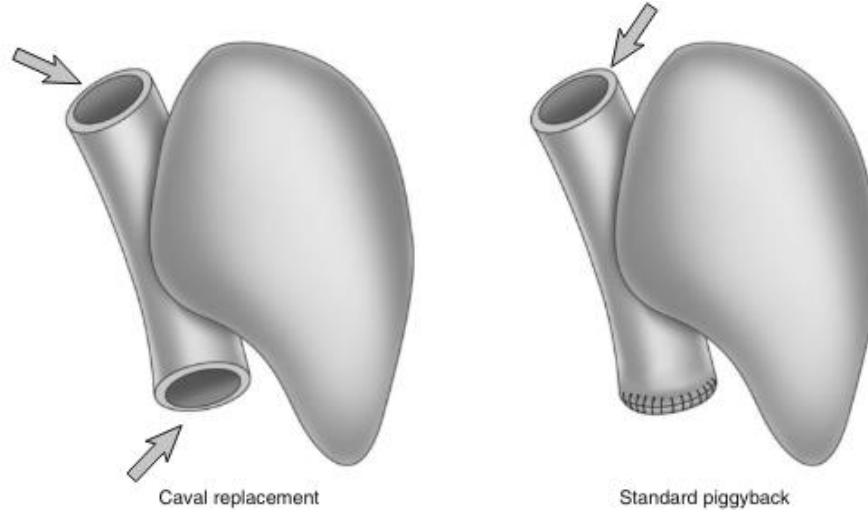
# Living Donor



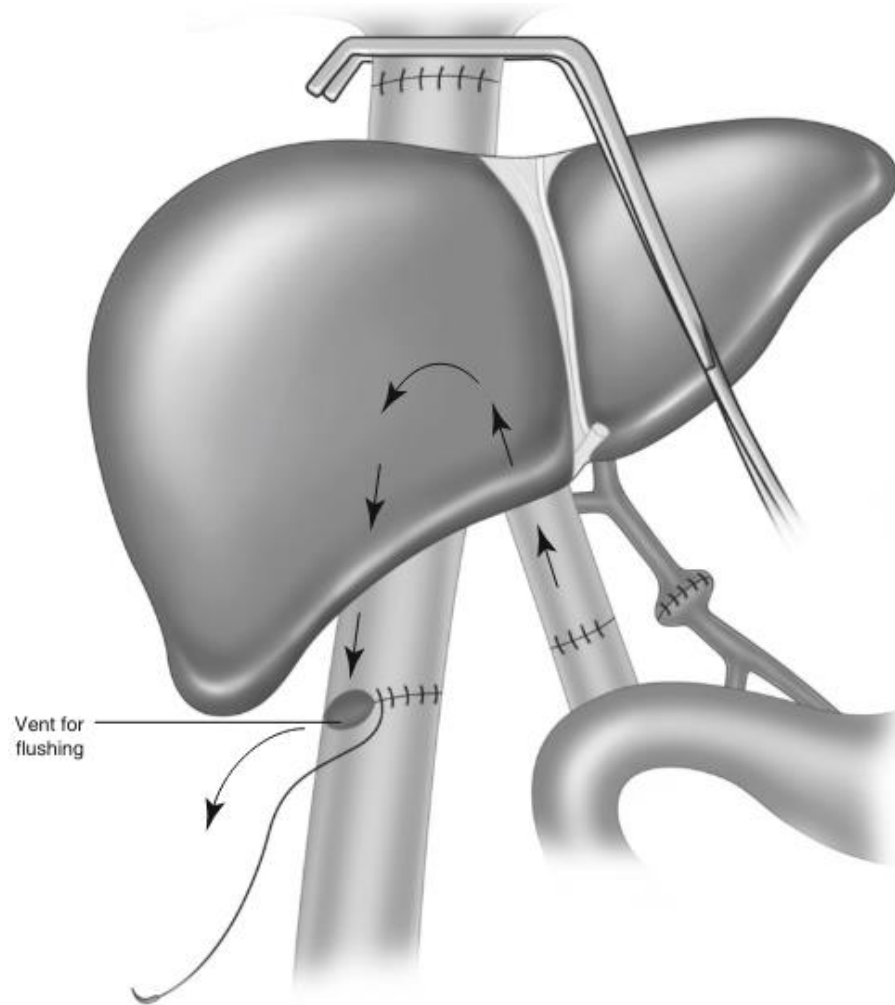
# Split Transplant



# Vena cava reconstruction

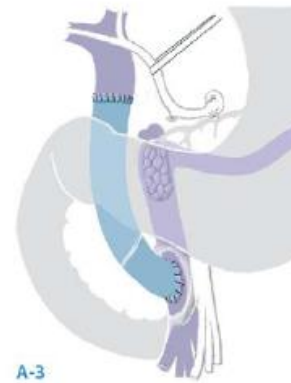
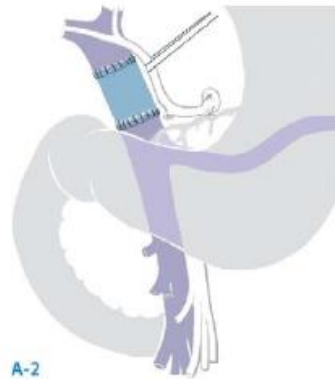
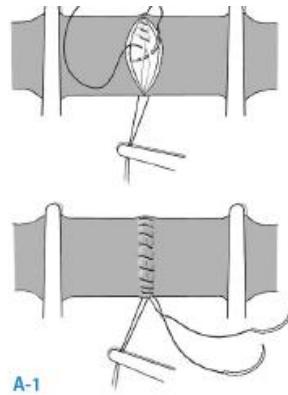


# Vena cava reconstruction

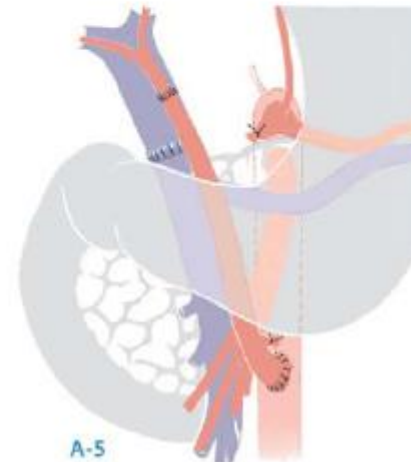
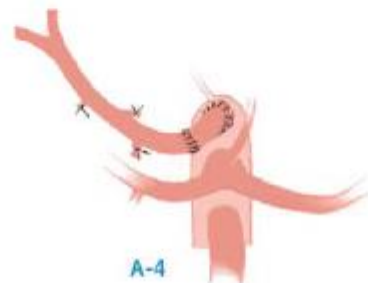
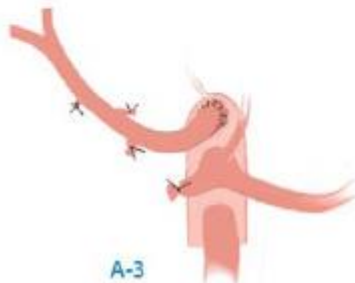
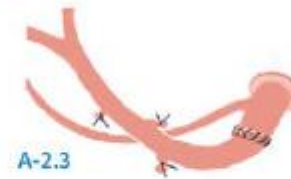
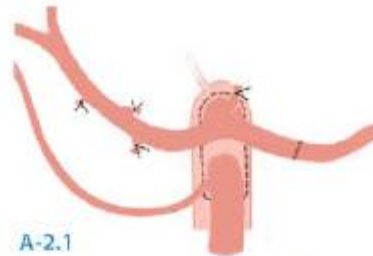




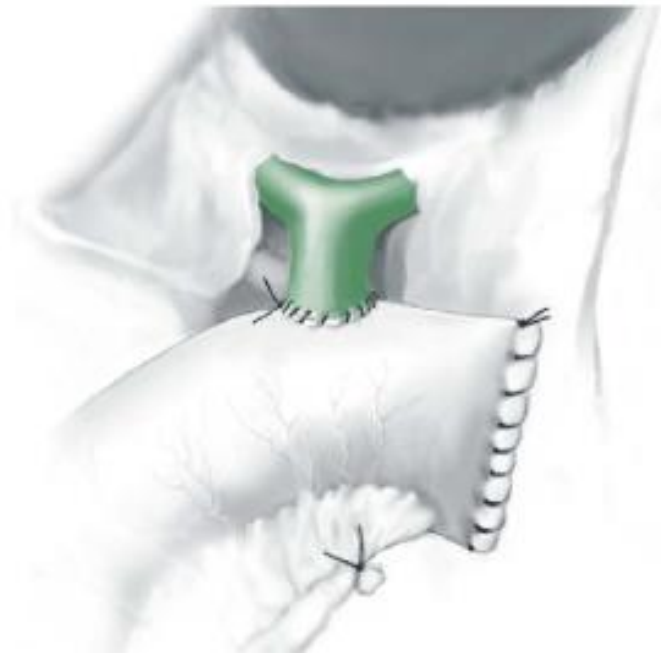
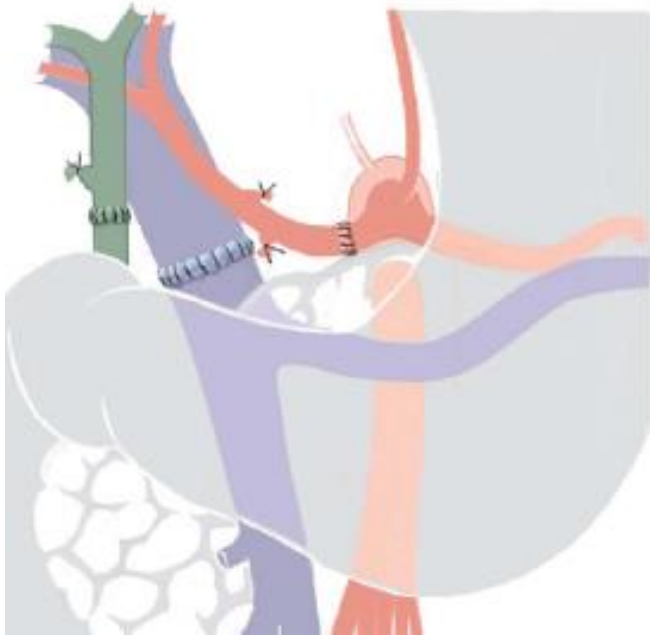
# Portal vein



# Hepatic Artery



# Bile ducts



# Postop Course



# Potential complications

- Bleeding
- Thrombosis
- Biliary
- Wound
- Infections
- Rejection



# Postop Course



# Postop Course



# Postop Course





# Postop Course



# Postop Course

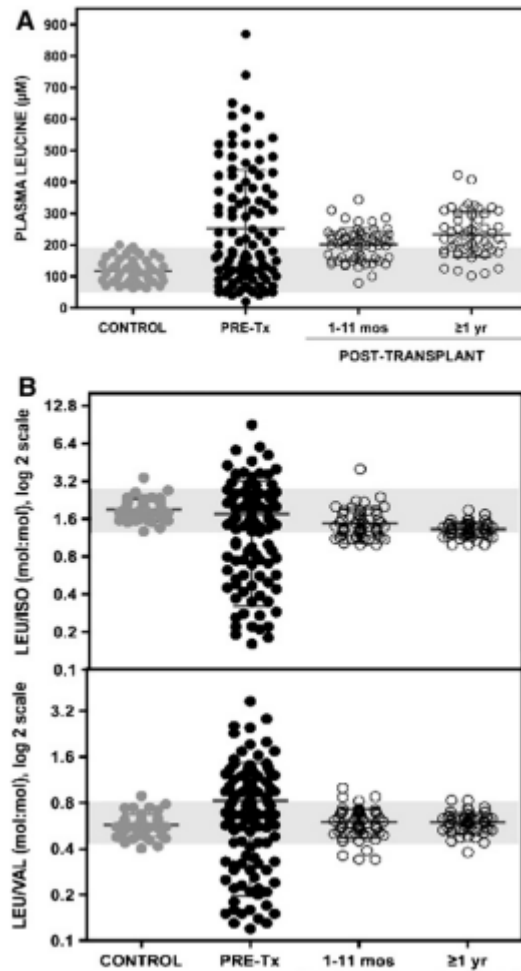


# Postop Course



# Liver Transplantation for Classical Maple Syrup Urine Disease: Long-Term Follow-Up in 37 Patients and Comparative United Network for Organ Sharing Experience

George V. Mazariegos, MD<sup>\*1</sup>, D. Holmes Morton, MD<sup>2</sup>, Rakesh Sindhi, MD<sup>1</sup>, Kyle Soltys, MD<sup>1</sup>, Navdeep Nayyar, MD<sup>1</sup>, Geoffrey Bond, MD<sup>1</sup>, Diana Shellmer, PhD<sup>5</sup>, Benjamin Shneider, MD<sup>5</sup>, Jerry Vockley, MD<sup>6</sup>, and Kevin A. Strauss, MD<sup>\*,2,3,4</sup>



**Figure 1. A,** Pooled plasma leucine values in control subjects (gray circles) and patients with MSUD before (black circles) and 1-11 months and  $\geq 1$  year after liver transplantation (open circles). Shaded area represents mean  $\pm 2$  SD leucine values in 51 normal children. A single exceptional value of 2170  $\mu\text{M}$  (not shown) was seen in a child who developed severe dehydration at 55 months posttransplantation. **B,** Plasma molar ratios of leucine to isoleucine (upper panel) and leucine to valine (lower panel) showing intact regulation of BCKDH activity after transplant (open circles). Shaded areas represent mean  $\pm 2$  SD molar ratios in normal children.

. (J Pediatr 2012;160:116-21).

# Liver Transplantation for Classical Maple Syrup Urine Disease: Long-Term Follow-Up in 37 Patients and Comparative United Network for Organ Sharing Experience

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**Table.** Perioperative and postoperative complications in 37 patients

	Number	Percent
Postsurgical interventions		
Delayed wound closure	10	27
Ventral hernia repair	4	11
Gastrocutaneous fistula closure*	2	5
Exploratory laparotomy:		
Hepatic artery thrombosis with successful revision	2	5
Hepatic artery revision or graft revision	3	8
Intra-abdominal bleeding	1	3
Partial small bowel obstruction	1	3
Pleurocentesis	2	5
Chest tube drainage	2	5
Bronchoscopy	1	3
Medical complications		
Acute rejection <sup>†</sup>	15	40
Epstein-Barr virus disease	2	5
Cytomegalovirus disease	1	3
Posttransplantation lymphoproliferative disease <sup>‡</sup>	1	3

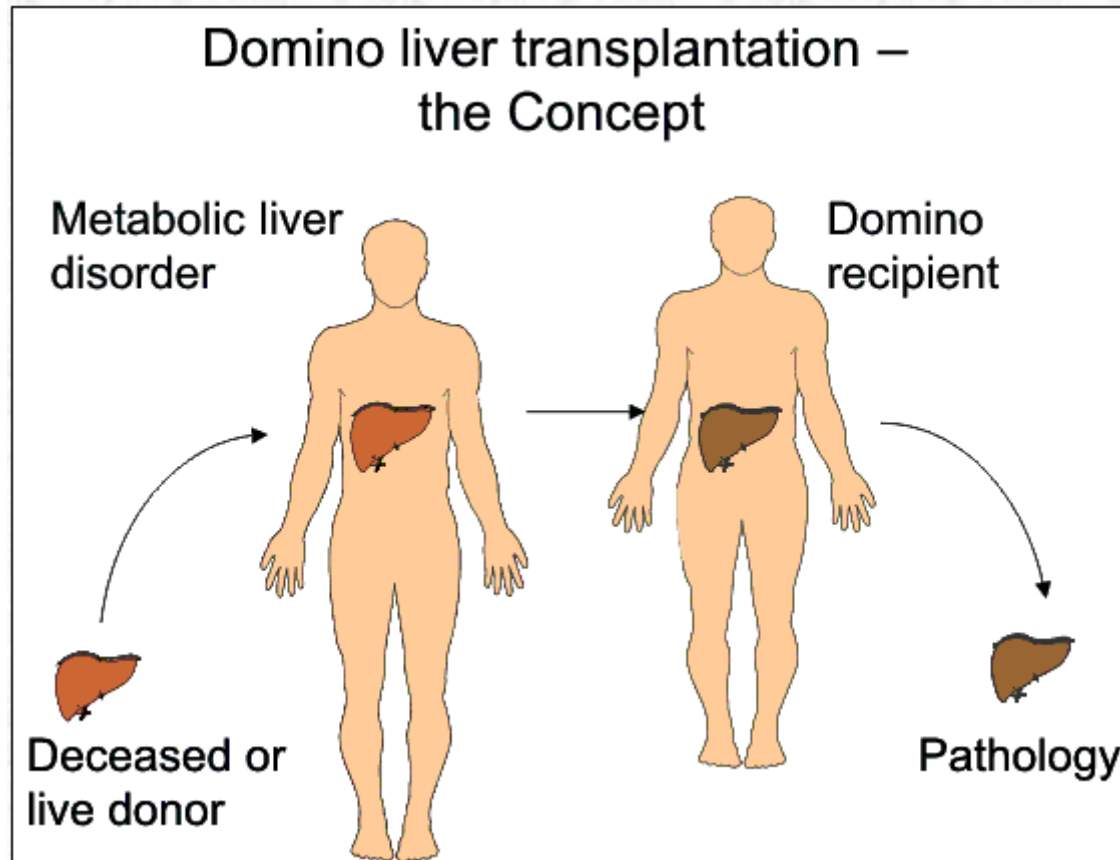
\*At previous gastrostomy tube sites.

<sup>†</sup>Antibody therapy for steroid-resistant rejection in 3/15 (8% of all patients).

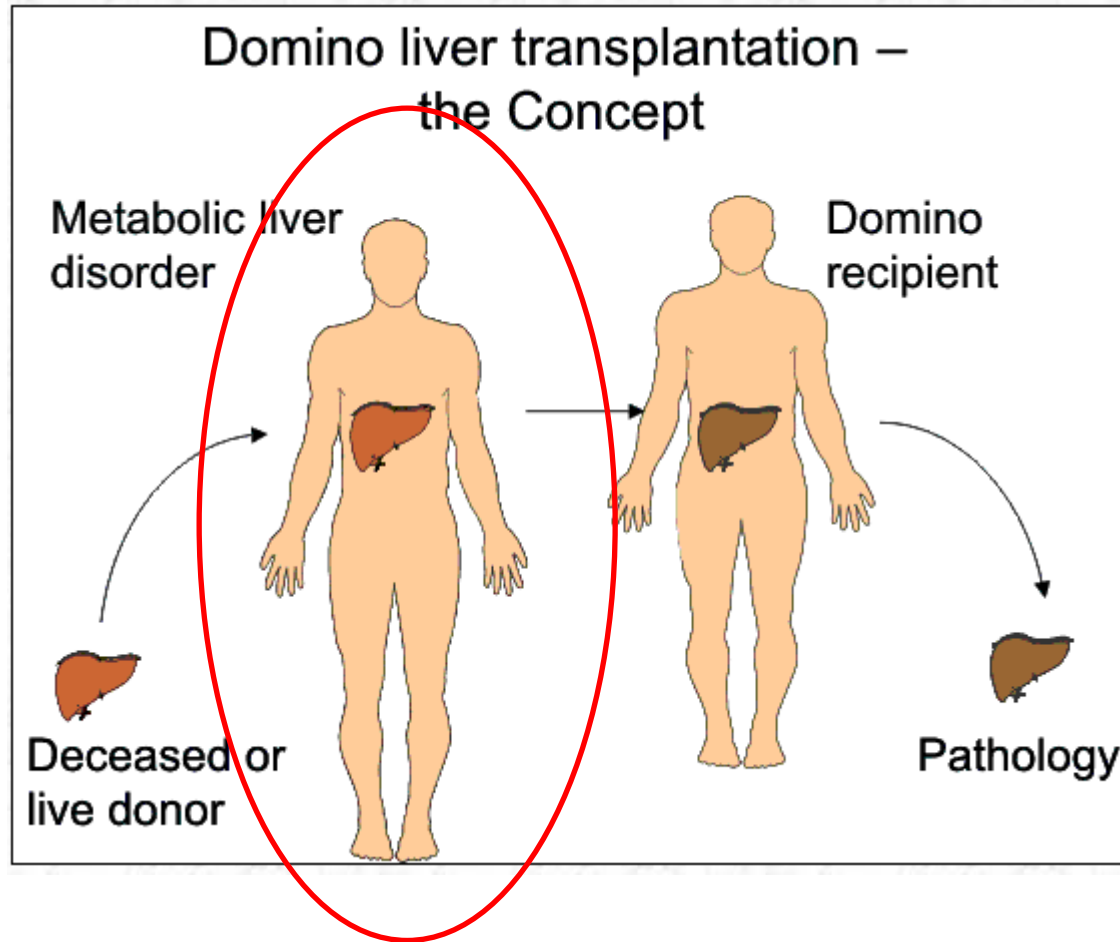
<sup>‡</sup>Intestinal posttransplantation lymphoproliferative disease developed in 1 patient who underwent transplantation at another center; it resolved with transient withdrawal of immunosuppression, and the patient has been disease-free for 12 years.

. (*J Pediatr* 2012;160:116-21).

# Domino Transplant



# Domino Transplant



# Domino liver transplantation: Expanding the liver donor pool to the pediatric recipient

[No Title]

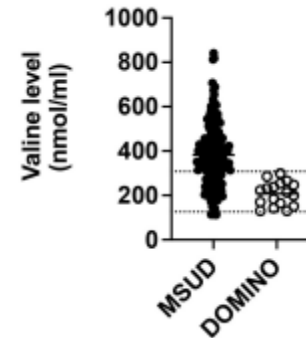
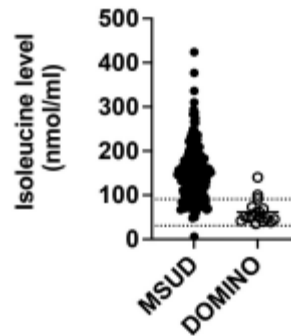
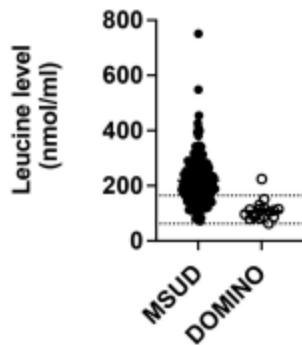
**TABLE 1** Clinical characteristics of domino liver donors and recipient pairs

Domino LT recipient	Pre-LT diagnosis	Waiting time (days)	Age (years)/weight (kg) at the time of domino LT			MELD/PELD at TX	Postoperative complications within 3 years after LT (vascular/biliary)
			Domino recipient	MSUD donor	GRWR		
Pediatric 001	Secondary sclerosing cholangitis	120	1/8.07	0.62/9.08	4.1	35	None
Pediatric 002	Neonatal hepatitis	166	4.2/15.4	7.2/28.4	3.9	40	None
Pediatric 005	Alpha-1-antitrypsin deficiency	444	2.2/17.2	2.2/15.7	2.7	20	Biliary stricture
Pediatric 006	Biliary atresia	364	2.0/13.3	1.1/11.3	2.4	25	Right HV thrombosis
Adult 003	Hepatitis C	29	62.7/81.5	14.1/73	2.0	7	None
Adult 004	Hepatitis C/HCC	164	67.9/48.8	10.2/45	2.0	25	None
Adult 007	Primary sclerosing cholangitis	1354	36.8/45.5	6.8/35.6	1.8	13	None
Adult 008	Alcoholic cirrhosis	175	55.3/63.5	6.2/32.1	0.9	17	None
Adult 009	Primary sclerosing cholangitis	618	49.1/53.3	14.5/47.2	1.5	22	None
Adult 010	Hepatitis C	776	68.2/50.4	16.5/86.1	3.5	15	HA stenosis biliary stricture



# Domino liver transplantation: Expanding the liver donor pool to the pediatric recipient

[No Title]



# Domino Transplant

- Deceased / LD LT
- Logistics
- Informed Consent
- Recipients; Adult / pediatric



# Liver Transplant for MSUD

- Quality of Life
- Risks and benefits
- Multiple surgical options
- Domino tx

# Thank you !!

