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BRIDGING LIFE & LIVER:
UNDERSTANDING PEDIATRIC LIVER TRANSPLANTATION

SPEAKER
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CHAIRPERSONS
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Dr Raghvendra Singh

DESIGNATION	**Professor (Pediatrics)** **Incharge Pediatric Gastroenterology, Nutritional Rehabilitation Centre and GI Endoscopy Lab**
CURRENT AFFILIATION	Maulana Azad Medical College and Lok Nayak Hospital, Delhi
ACHIEVEMENTS	Fellow, Pediatric Gastroenterology Hepatology and Nutrition (2023-24), SickKids, University of Toronto, Canada Treasurer 2025-26, Pediatric and Adolescent Nutrition Society (IAP Nutrition Chapter)

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Dr Abhideep Chaudhary

Vice chairman & HOD

BLK Max Centre for HPB Surgery & Liver Transplantation

President LTSi –elect

(Liver Transplant Society of India)

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Liver transplant in children

Dr Abhideep Chaudhary
Vice chairman & HOD
HPB Surgery and LT

Dr Ravi Bharadwaj
Sr Consultant
Pediatric gastroenterology

Dr Raghvenra Singh
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Incharge Pediatric Gastroenterology
MAMC

Dr Praveen Kumar
Pediatric Gastroenterologist
Professor & Head
Kalawati Saran Children
Hospital.

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Liver transplant

Human liver transplantation - first attempted - 1963 - EHBA

Outcomes improved following the introduction of cyclosporine in 1980

The first liver transplant in children in India was **performed in 1998 at Apollo Hospital Delhi**

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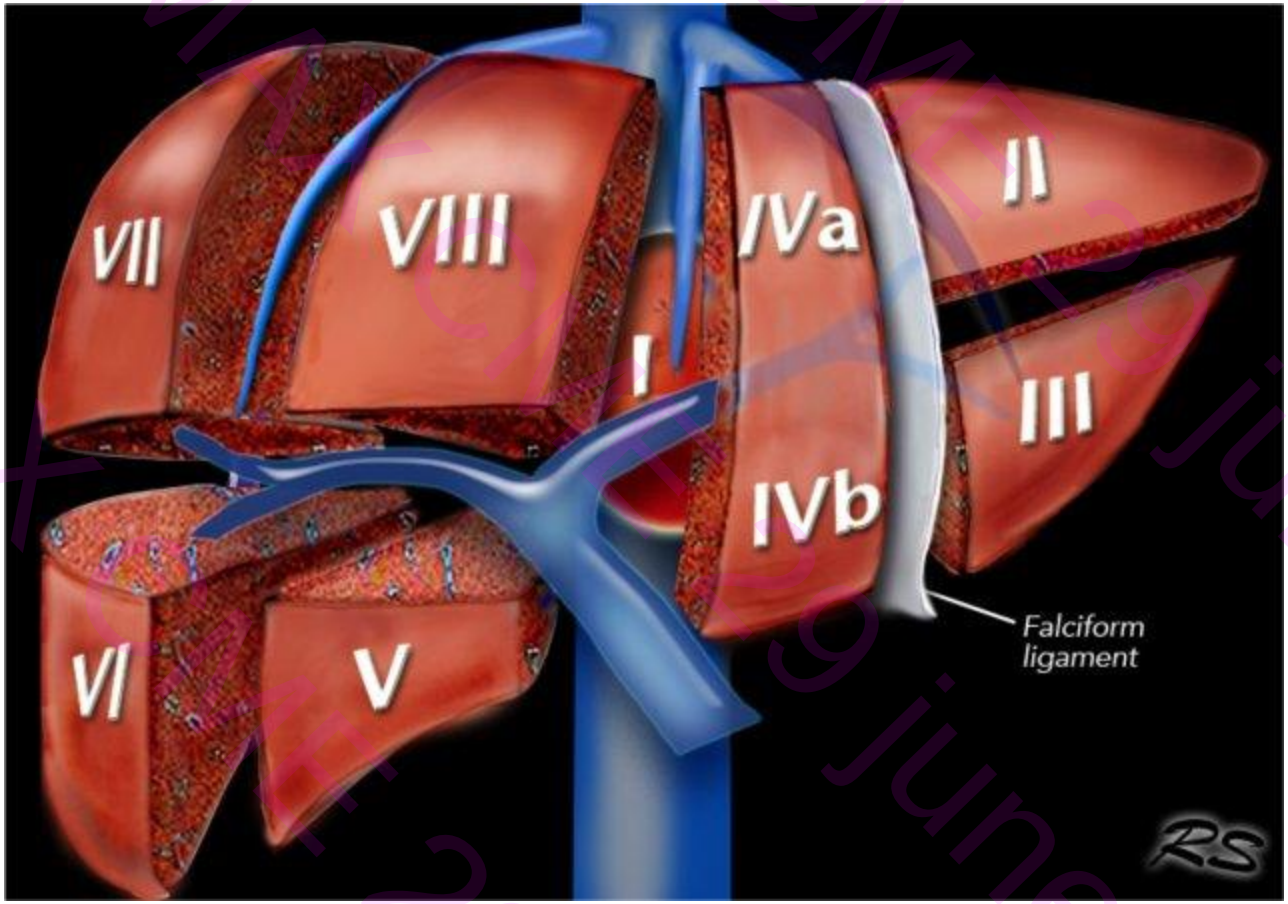
Couinaud segments



For children – typically left lateral segment 2, 3 or lateral Seg 2,3,4

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Imp indications for LT

Chronic Cholestatic Disease 54.3%
Biliary atresia 41.1%
Alagille syndrome 2.9%
Primary sclerosing cholangitis 2.7%
TPN-induced cholestasis 1.8%
Progressive intrahepatic cholestasis 1.5%
Idiopathic cholestasis 1.1%
Neonatal hepatitis 1.0%
Biliary cirrhosis, other cholestatic diseases 2.2%
Acute Liver Failure 13.8%
Cirrhosis 6.7%
Autoimmune hepatitis with cirrhosis 2.9%
Neonatal hepatitis cirrhosis 0.5%
Metabolic Disease 14.4%
α1-Antitrypsin deficiency 3.0%
Urea cycle defects 2.4%
Cystic fibrosis 1.6%
Wilson's disease 1.2%
Tyrosinemia 1.0%
Primary hyperoxaluria 0.7%
Crigler-Najjar syndrome 0.7%
Glycogen storage disease 0.7%
Neonatal hemochromatosis 0.5%
Inborn error in bile acid metabolism 0.1%
Primary Hepatic Malignancy 6.2%
Hepatoblastoma 4.2%
Other 2%
Other 4.7%
Congenital hepatic fibrosis 1%
Budd-Chiari syndrome 0.4%
Toxicity 0.7%

Transplantation 4 th edition

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A good liver donor is someone

Healthy,
Close relative of the recipient
Has a compatible blood type and body size

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Criteria for live donation of a liver:

Must be in good physical and mental health

Must be between the ages of 18 and 60

Must have a body mass index (BMI) that is less than 32

Must have a compatible blood type with the recipient

Must be free from the following:

- Significant organ diseases (i.e., heart disease, kidney disease, etc.)
- Ongoing malignancy (cancer)
- Hepatitis
- Active or chronic infections
- Active substance abuse

Finally, the donation of any organ by a living person must be completely voluntary.

Donors should be free from any pressure or guilt associated with the donation and cannot be paid for their donation.

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Donor time line

Admission -1 (1 day prior to LT)

OT time typically between 6-10 hrs

On table extubation

ICU stay for around 2 days

Discharge on 7th day

Can resume normal usual activites by 1 month post OT

Can resume strenuous activities typically after 3 months

Approx mortality 1 in 2500 (No donor death at our centre)

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GENERAL INDICATIONS FOR LIVER TRANSPLANTATION IN CHILDREN

- Primary Liver Disease That leads to Hepatic Insufficiency
- Acute Liver Failure
- Liver Transplantation as Primary Therapy for Inborn Errors of Metabolism
- Secondary Liver Disease
- Primary Hepatic Malignancy

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Indications for liver transplant (EHBA)

Once the opportunity of KPE is missed > 100 days

Primary failure of the Kasai

- **At 3 months or more post Kasai**
- **TB >6 mg/dl : prompt referral for LT evaluation**
- **TB 2-6 mg/dL : LT evaluation should be considered**

Refractory growth failure

Recurrent cholangitis

Complications of PHTN (POPH, HPS, ascites, recurrent bleeding)

Progressive liver dysfunction

Squires RH et al, Hepatology. 2014;60(1):362.
Jiang CB et al, Eur J Pediatr. 2003;162(9):603.
Barshes NR et al, Liver Transpl. 2005;11(10):1193.

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ALF- all 3 criteria

Acute onset – hepatic failure within eight weeks of onset of clinical liver disease
no previous evidence of chronic liver disease

Biochemical evidence of acute liver injury (one or both):

- **Hepatocellular injury – AST or ALT) >100 IU/L (unless explained by myopathy)**
- **Biliary dysfunction – TB >5 mg/dL , DB >2 mg/dL and/or GGT >100**

Coagulopathy – Persists after inj vitamin k administration

- **PT≥15 seconds or international normalized ratio (INR) ≥1.5 with evidence of hepatic encephalopathy**
- **PT ≥20 seconds or INR ≥2.0, with or without encephalopathy.**

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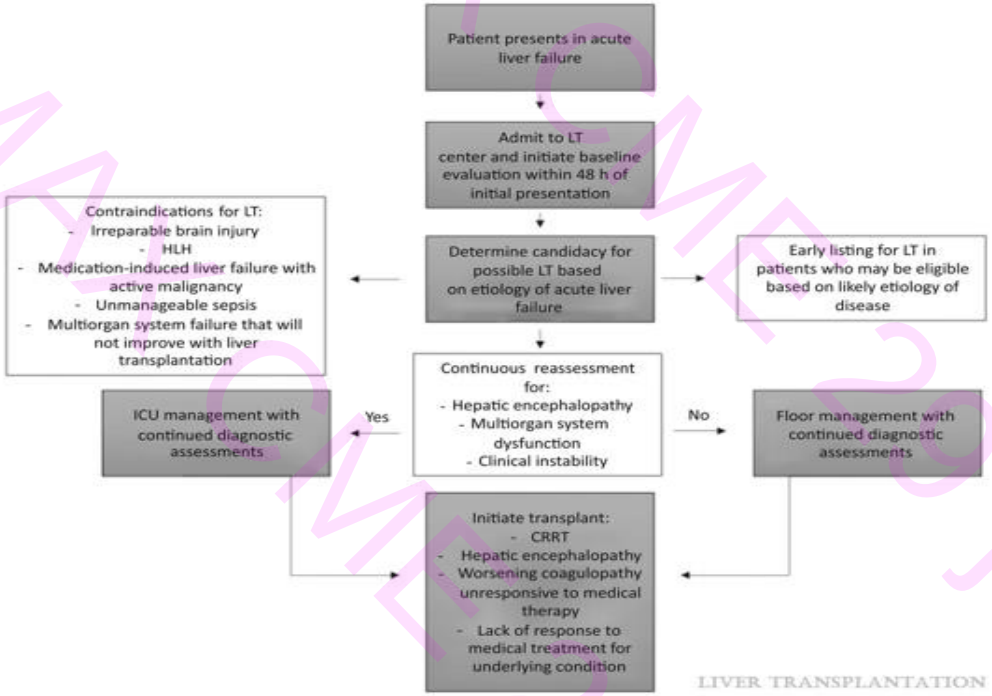
LT in PALF

FIGURE 1



Pediatric acute liver failure: Reexamining key clinical features, current management, and research prospects

Ascher Bartlett, Johanna M.; Yanni, George; Kwon, Yong; Emamaullee, Juliet

Liver Transplantation28(11):1776-1784, November 2022.

doi: 10.1002/lt.26500

Proposed approach to managing patients with PALF.

AASLD AMERICAN ASSOCIATION FOR THE STUDY OF LIVER DISEASES

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Liver transplant in PALF

Of note, the degree of coagulopathy has not been shown to correlate with hepatic encephalopathy.

The PALFSG has demonstrated that 25% of patients who had Grade 3 or 4 hepatic encephalopathy and required intensive care had mild coagulopathy with INR <2.0, yet patients with this degree of encephalopathy (Grade 3–4) also demonstrated the highest rates of mortality.

All patients with acute liver failure require close monitoring and daily assessment of overall mental status for this reason.

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PALF

The Liver Injury Unit score has been developed specifically for PALF and includes factors for peak total bilirubin, PT or INR, and ammonia

Sensitivity and specificity were 74 and 80 percent

LIU score <209- low risk

LIU score >370- high risk

NWI (New Wilson Index >11=LT)

PELD

PELD Score = 0.480 x ln(bilirubin in mg/dL) + 1.857 x ln(INR) - 0.687 x ln(albumin in g/dL) + 0.436 if the patient is <1 year old + 0.667 if there growth failure

LIU score = 3.507 × peak total bilirubin (mg/dL) + 45.51 × peak INR + 0.254 × peak ammonia (µmol/L

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Poor prognostic indicators for PALF include

Younger than 1 year at the time of presentation

Presence of Grade 4 encephalopathy

The Liver Injury Unit (LIU) score; is able to predict death or LT by 4 weeks of patient presentation with an area under the curve of 88.5%–90.5%
Use in the clinical setting is limited by a reliance on peak laboratory values

The King's College model incorporates the presence of hepatic encephalopathy, which is an unreliable feature in pediatrics

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Poor prognostic indicators for PALF include

Patients most likely to be listed

higher INR (median, 3.0), total bilirubin (median, 15 mg/dl), lactate (median, 2.8 mmol/L), ammonia (63 µmol/L), and lower liver enzymes (alanine aminotransferase, 1635 IU/L), boys, inotrope, MV, Indeterminate PALF

Resolution of disease without transplant was more likely in patients without encephalopathy

Negative impact of young age on PALF outcomes

Bhatt H et al , Curr Pediatr Rep. 2018;6:246–57.
Jain V, Dhawan A.. Liver Transpl. 2016;22:1418–30.
Squires JE et al. Hepatology. 2018;68:2338–47.

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Liver support

Extracorporeal liver support systems

Artificial liver support, including the membrane-adsorbent recirculating system and plasma exchange (with or without hemodialysis), and bioartificial liver support

Plasmapheresis or plasma exchange

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Liver Transplantation as Primary Therapy for Inborn Errors of Metabolism

Special mention- Liver transplantation can benefit children with inborn errors of metabolism that do not injure the liver, the principal goal of treatment being to correct the metabolic error

Urea cycle defects

Crigler-Najjar syndrome,

Homozygous familial hypercholesterolemia

Primary hyperoxaluria

The decision of whether to perform liver transplantation depends on knowing

- it will correct the metabolic defect
- there is no effective alternative therapy,
- patient has not experienced irreversible complications.

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UCD

The decision-making process is different for urea cycle defects, which result in hyperammonemia and brain damage.

Despite advances in medical management, severe defects such as ornithine transcarbamylase (OTC) deficiency in males still have a very poor outcome.

OTC deficiency is an X-linked disease.

Boys with OTC deficiency should be considered for transplantation immediately upon making the diagnosis

Successful transplantation corrects the metabolic defect but cannot undo preexisting brain damage.

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Secondary Liver Disease

Cystic fibrosis and biliary cirrhosis

Sclerosing cholangitis secondary to Langerhans cell histiocytosis

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Primary Hepatic Malignancy

Hepatoblastoma:

Rescue transplants" carry a much worse prognosis than tumors treated by primary transplantation

Liver transplants that are done after a tumor has recurred in the liver carry a nonrecurrence rate of 20% to 30% in comparison to rates of greater than 90% for primary transplants.

Transplantation should be considered only if complete resection is not possible

Hepatocellular carcinoma is extremely rare in children outside the context of

Metabolic liver disease
Tyrosinemia

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Typical Pre LT Course

Assessing the etiology and need

Nutritional optimization (Vit A, D, E, K, MCT Oil, Multivitamins and Ca)

Management of complications- PHT , Ascites, HPS, Sepsis, Infection

Donor and recipient work up

Clearances

Typically takes around 1-2 wk depending on urgency

Admitted on -1 (1 day prior to LT)

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Typical Posttransplant Course

"Fast-track" approach entailing extubation inthe operating room

ICU stay of 3 to 5 days

Oral feedings around 2nd -3rd day

Median length of hospital stay - about 15 to 20 days

Laboratory test results are checked on a weekly basis after discharge

Medications are tapered according to center protocol, with most patients on calcineurin monotherapy by 1 year after transplantation

Lifelong immunosuppression is the standard of care at most centers, but this might change for select groups of recipients that achieve operational tolerance with stable histological characteristics and graft function

Late complications such as biliary strictures, vascular occlusions

Live vaccines are prohibited until the patient is on monotherapy for at least 6 months

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GENERAL CONTRAINDICATIONS TO LIVER TRANSPLANTATION

- Secondary organ failure
- Severe pulmonary hypertension
- Severe portopulmonary hypertension
- Presence of disease that is expected to recur after therapy
- Metastatic carcinoma

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Survival and Quality of Life in Children

The 5-, 10-, 15-, 20-, and 25-year survival rates for pediatric liver transplant recipients are 85.0%, 84.7%, 84.2%, and 80.8%, respectively

The 5-, 10-, 15-, 20-, and 25-year graft survival rates for pediatric liver transplant recipients are 81.6%, 77.6%, 76.3%, 75.0%, and 75.0%, respectively

The projected 20-year survival rate for pediatric liver transplant recipients transplanted between 2007 and 2018 is 84.0%, and the projected 30-year survival rate is 80.1%.

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Survival and Quality of Life in Children

	Survival	Graft survival	Projected survival
5 year	80-85	78-81.6	
10 year	79-84.7	75-77.6	
15 year	79-84.2	75-76.3	
20 year	75-81	73-75	84
25 year	73-80.8	71-75	

Multiple single centre studies

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2019	1
2020	1
2021	6
2022	12
2023	17
2024	14
2025	7
total	58

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male	33
female	25

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Age	N.
<1 yr	13
1-5	14
>5	31

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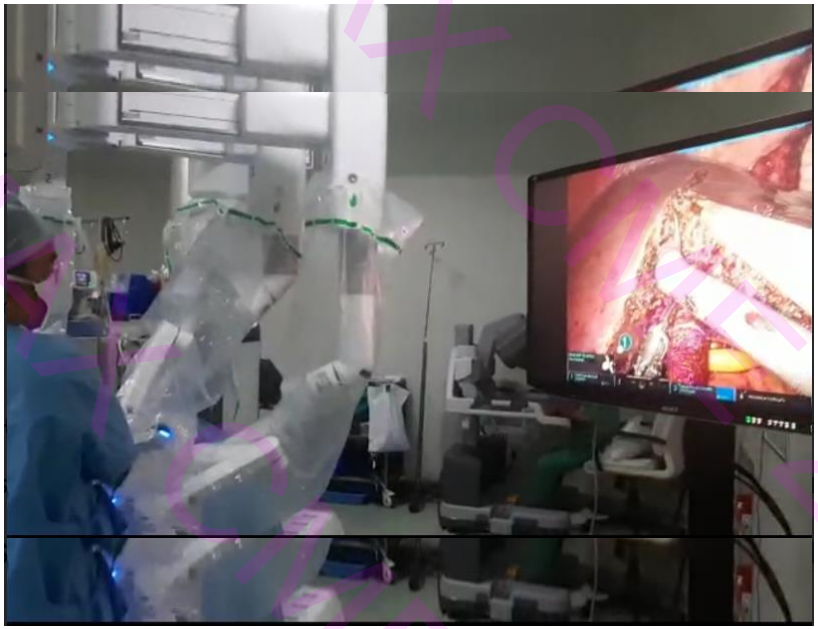
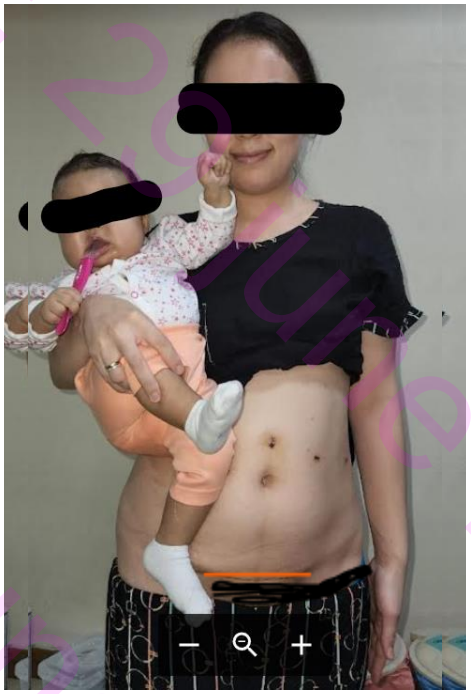
EHBA (pre and post kasai)	17
PFIC	10
Wilson	5
AIH	8
BCS	2
CDC	1
GSD/Metabolic	3
ALF Hepatoblastoma Cryptogenic	5 2 5

ROBOTIC DONOR LEFT LATERAL HEPATECTOMY





Total robotic Donor	26
Robot assisted	17
Complete Robotic	9
Left Lateral graft	**3**

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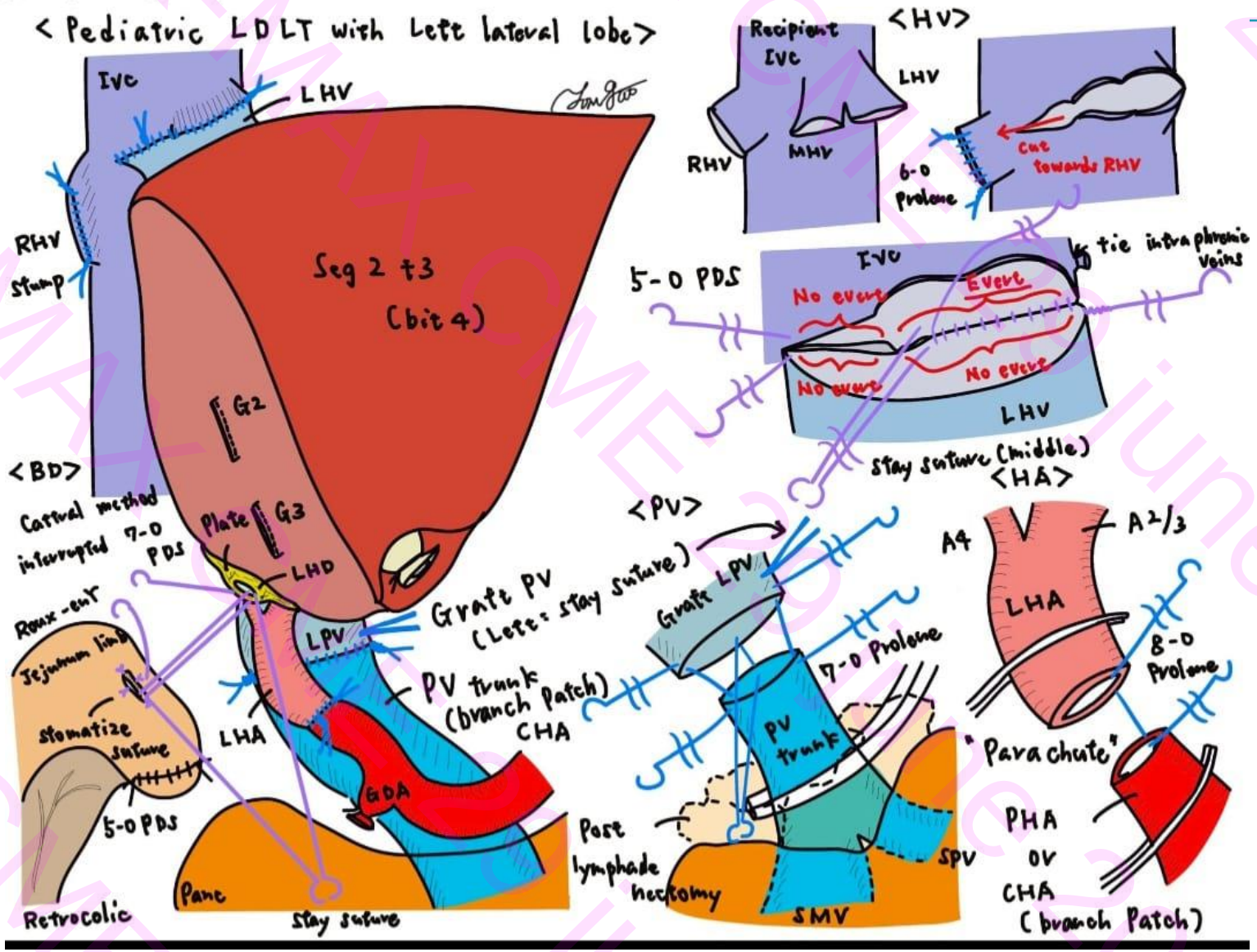

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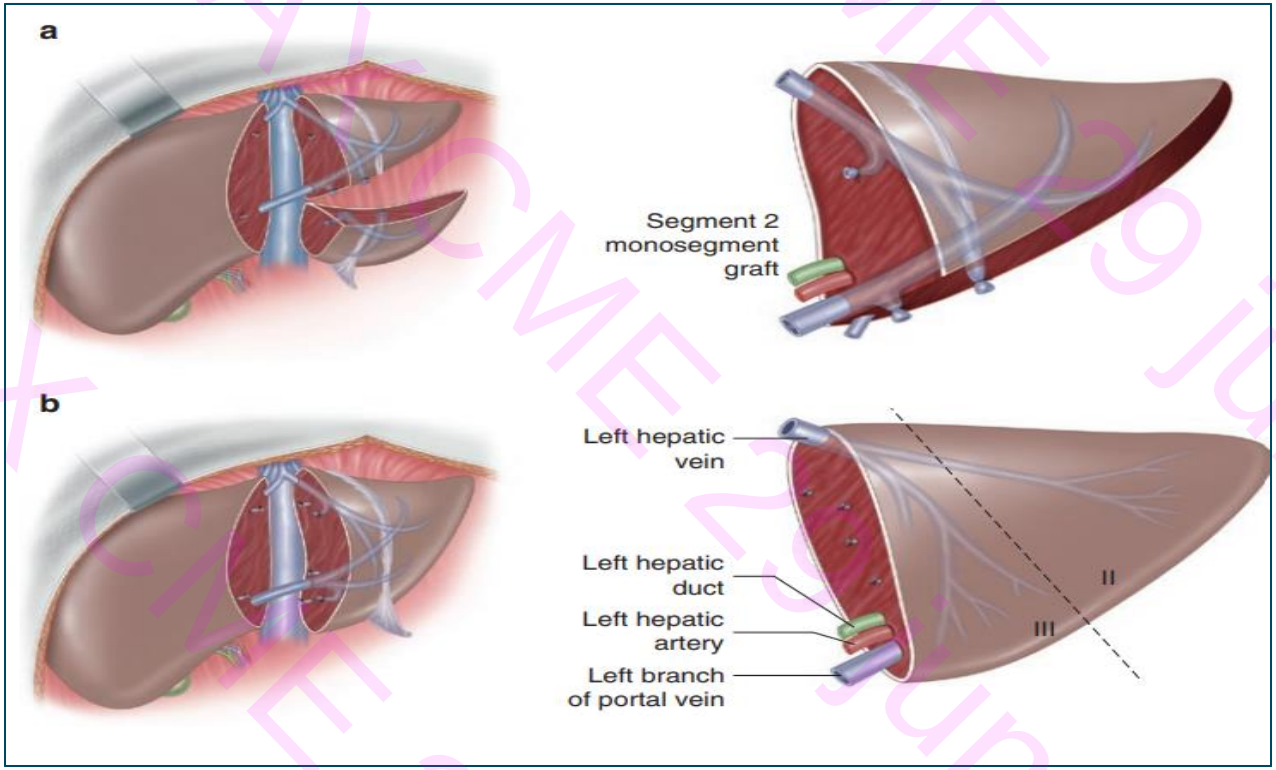
Left lateral segment hyper-reduction technique



Child wt. <5kg

Graft to recipient weight ratio >4 %

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PALF

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PALF

Pre LT

Day 3 –post LT

Day 10 –post LT

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Surviving the odds!

ALF

Hepatoblastoma

AILF, Tx, HAT,
ReTransplant

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From yellow to white

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Growth/Height

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Youngest at 4 months

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I am no more an infant

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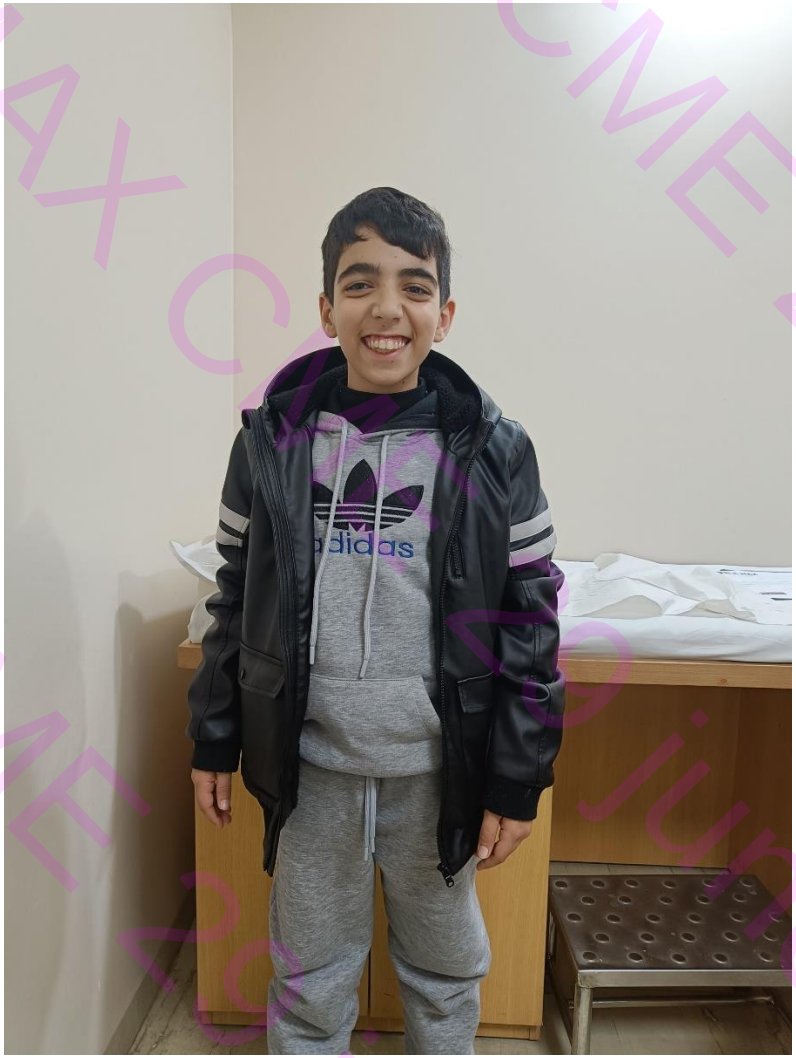
Sometimes you get more than you ask



Decompensated liver disease , Wilson and mild neurological impairment

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Story of trust
from being scared to sporty

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Birthday in LT

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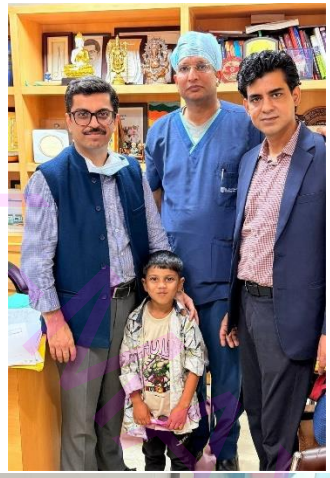

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Severe HPS

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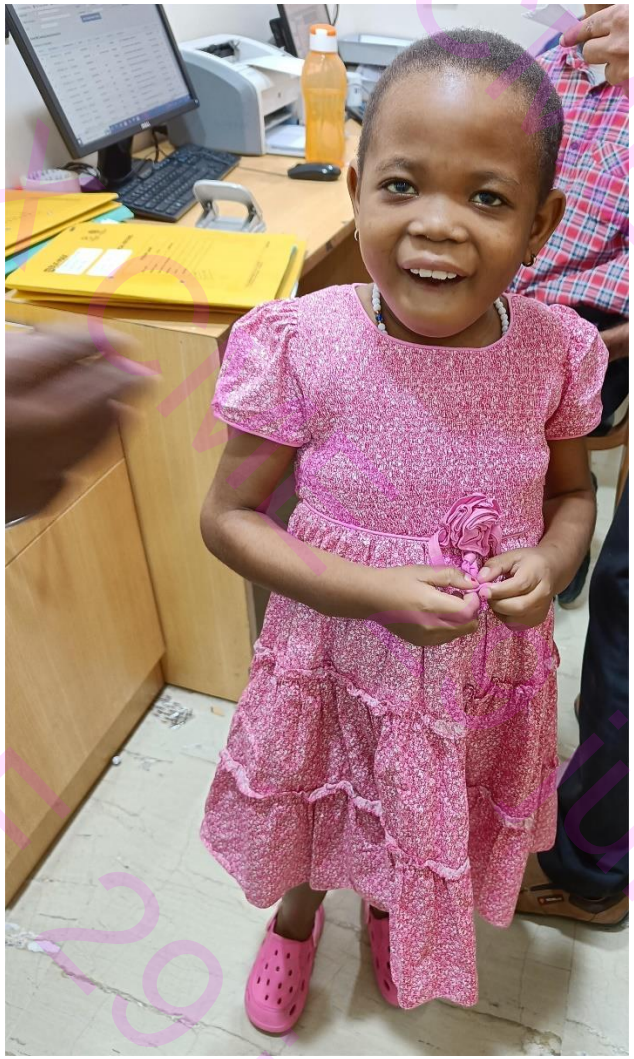
I don’t need a transplant



And I don’t need a wheel chair

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ACLF, sepsis, dystonia ,

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Difficulty in standing to learning Indian dance now

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She is now learning to walk

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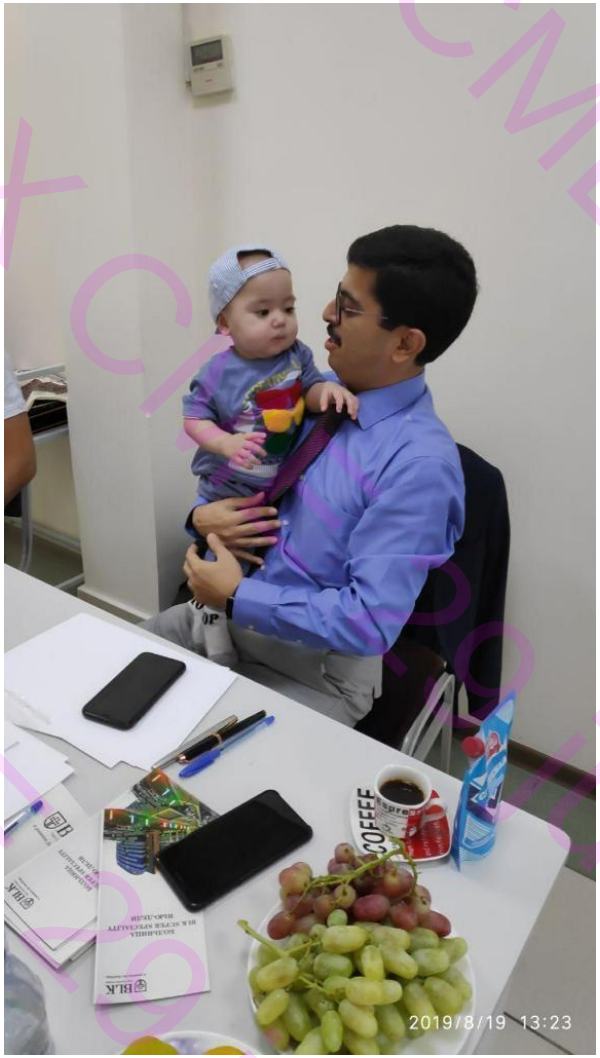
Now you know who calls the shots here



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Thank you