

Pediatric Liver Intensive Care

Naresh Shanmugam
Anil Dhawan
Editors

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To my mentors Prof. Anil Dhawan and Prof. Mohamed Rela who inspired me in every aspect of life and made me look beyond the horizon.

Naresh Shanmugam

Preface

Pediatric hepatology has evolved into a distinct subspecialty, and pediatric liver transplant care has become an integral part of it. Intensive care management of children with liver disease requires the coordination of a multidisciplinary team as decisions have to be made regarding the need for transplantation, strategies to be used during transplantation, and post-transplant care. Though there are several standard textbooks available on pediatric hepatology, this manual on *Pediatric Liver Intensive Care* is unique as it is written from an intensivist perspective that helps in the management of common liver problems to complex hepatobiliary/peri-transplant care. This manual has been written in a ready reckoner format that could be used at the bedside. The editors have taken into consideration a wide variety of readers including transplant surgeons and anesthesiologists who deal with pediatric transplants and have incorporated management protocols, common pediatric drug dosages, etc. for their easy use. The chapters have been written by experienced authors who have both intensive care and pediatric hepatology knowledge from high volume pediatric liver transplant centers. Some of the authors across specialties such as radiology, cardiology, and anesthesia have shared their insights regarding decision-making and management principles from their perspective.

The practical knowledge of the authors in the field of pediatric hepatology is reflected by “practical tips” and “caution alert” provided by them in each chapter. We sincerely hope that this manual would bridge the knowledge gap between various specialists, such as pediatrician, intensivist, metabolic consultant, transplant surgeon, and anesthesiologist, and help in the management of children with liver disease.

Chennai, India
London, UK

Naresh Shanmugam
Anil Dhawan

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About the Editors

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Anil Dhawan is a consultant in pediatric hepatology at King's College Hospital, London. He is the head of the Liver GI and Nutrition Centre and MowatLabs and the Corporate Medical Director of the Variety Children's Hospital, London. Prof. Dhawan is the founder and head of the Basic Science Laboratories (Mowat Labs) at the Institute of Liver Studies, King's College Hospital, London.

He has held board-level appointments at the European Society of Pediatric Gastroenterology, Hepatology and Nutrition, International Liver Transplantation Society, and Cell Transplantation Society. He was the president of the Cell Transplant and Regenerative Medicine Society. He is on the editorial board of several journals and has published more than 300 peer-reviewed articles in the field of hepatology and hepatocyte transplantation and has edited four textbooks on liver disease in children and hepatocyte transplantation. He is a regular invited speaker at prestigious adult and pediatric liver meetings across the world.

Abbreviations

99mTc-MAA	Technetium-99m-labeled macroaggregated albumin
ABG	Arterial blood gas analysis
ACD	Acid citrate dextrose
ACR	Acute cellular rejection
ADA	Adenosine deaminase
AED	Antiepileptic drug
AGT	Alanine: glyoxylate aminotransferase
AKI	Acute kidney injury
ALF	Acute liver failure
ALT	Alanine aminotransferase
APTR	Activated partial thromboplastin ratio
aPTT	Activated partial thromboplastin time
ASD	Atrial septal defect
ASFA	American Society for Apheresis
ATG	Anti-thymocyte globulin
ATN	Acute tubular necrosis
BA	Biliary atresia
BASM	Biliary atresia splenic malformation
BCAA	Branched chain amino acids
BCKD	Branched chain keto-acid dehydrogenase
BiPAP	Bi-level positive airway pressure
BNP	Brain natriuretic peptide
CBD	Common bile duct
CIT	Cold ischemic time
CLTK	Combined liver and kidney transplants
CNNA	Culture negative neutrocytic ascites
COG	Children Oncology Group
CPM	Central pontine myelinolysis
CPS	Carbamoyl phosphate synthase
CPT	Cryoprecipitate
CRS	Cytokine release syndrome
CT	Computed tomography
CVVH	Continuous veno-venous hemofiltration

CVVHD	Continuous veno-venous hemodiafiltration
DBD	Donation after brain death
DCD	Donation after cardiac death
DEXA	Dual energy X-ray absorptiometry
DIC	Disseminated intravascular coagulopathy
DiSA	Digital subtraction angiography
DO	Drain output
DSA	Donor-specific antibody
DSU	Doppler ultrasound
Dv	Diastolic velocity
ECG	Electrocardiography
ECHO	Echocardiography
ECV	Extracorporeal volume
EF	Ejection fraction
EFAs	Essential fatty acids
EMG	Electromyogram
ESLD	End-stage liver disease
ESPEN	European Society for Clinical Nutrition and Metabolism
EST	Endoscopic sclerotherapy
EVL	Endoscopic variceal ligation
FHVP	Free hepatic venous pressure
GGT	Gamma-glutamyltransferase
GI	Gastrointestinal
GIR	Glucose infusion rate
GRWR	Graft to recipient weight ratio
HA	Hepatic artery
HAS	Human albumin solution
HAS _t	Hepatic arterial stenosis
HAT	Hepatic artery thrombosis
HB	Hepatoblastoma
HCC	Hepatocellular carcinoma
HD	Hemodialysis
HE	Hepatic encephalopathy
HME	Heat moisture exchange
HOGA1	4-Hydroxy-2-oxoglutarate aldolase
HPS	Hepatopulmonary syndrome
HR	High-risk
HRS	Hepatorenal syndrome
HV	Hepatic veins
HVPG	Hepatic venous pressure gradient
ICA-AKI	International Club of Ascites–Acute Kidney Injury
ICP	Intracranial pressure
IMV	Inferior mesenteric vein
INR	International normalized ratio

KT	Kidney transplant
LAI	Liver Attenuation Index
LB	Liver biopsy
LCT	Long chain triglyceride
LDH	Lactate dehydrogenase
LDLT	Living donor liver transplantation
LOLA	L-ornithine L-aspartate
LPV	Left portal vein
LT	Liver transplantation
MAC	Mid-arm circumference
MB	Methylene blue
MCT	Medium chain triglyceride
MDT	Multidisciplinary team
MFD	Minimal fat diet
MHV	Middle hepatic vein
MMA	Methylmalonic acidemia
MMF	Mycophenolate mofetil
MNB	Monobacterial non-neutrocytic ascites
MRI	Magnetic resonance imaging
MSBOS	Maximum surgical blood ordering schedule
NAGS	<i>s</i> -acetyl glutamate synthase
NIV	Noninvasive ventilation
OGD	Esophagogastroduodenoscopy
OLT	Orthotopic liver transplantation
ORS	Oral rehydration solution
OTC	Ornithine transcarbamylase
PA	Propionic acidemia
PD	Peritoneal dialysis
PDA	Patent ductus arteriosus
PH	Primary hyperoxaluria
PHT	Portal hypertension
PI	Pulsatility index
PNF	Primary nonfunction
POD	Postoperative day
POPH	Portopulmonary syndrome
PPHT	Portopulmonary hypertension
PRES	Posterior reversible encephalopathy syndrome
PRETEXT	Presurgical pretreatment extent of disease
PSv	Peak systolic velocity
PT	Prothrombin time
PTBD	Percutaneous transhepatic balloon dilation
PV	Portal vein
PVT	Portal vein thrombosis
RDP	Random donor platelets
REE	Resting energy expenditure

RI	Resistive index
ROTEM	Rotational thromboelastography
RPF	Renal plasma flow
RRT	Renal replacement therapy
SAAG	Serum-ascites albumin gradient
SB	Sengstaken–Blakemore
SBP	Spontaneous bacterial peritonitis
SDP	Single donor platelets
SD-SST	Standard dose-short synacthen test
SIOPEL	Société Internationale d’Oncologie Pédiatrique– Epithelial Liver Tumor Study Group
SNS	Sympathetic nervous system
SR	Standard risk
SR	Sustained release
TEE	Total energy expenditure
TEG	Thromboelastogram
TEG	Thromboelastography
TIBC	Total iron-binding capacity
TIPS	Transjugular intrahepatic portosystemic shunt
TJ	Transjugular
tLPV	Transverse portion
TPE	Therapeutic plasma exchange
TST	Triceps skinfold thickness
TT	Tracheostomy tubes
UO	Urine output
USG	Ultrasonography
VAP	Ventilator-associated pneumonia
VSD	Ventricular septal defect
WHVP	Wedge hepatic venous pressure
WIT	Warm ischemic time



Liver Anatomy for Pediatric Intensivist

1

Mettu Srinivas Reddy

Children have a larger liver in relation to their body weight as compared to adults. While the liver to body weight ratio in adults is around 2%, it is around 4% in infants. The segmental anatomy in children is very similar to that in adults.

1.1 Attachments of the Liver

The liver is located in the right subphrenic space and kept in position through its attachment to the cava and the peritoneal folds or ligaments which continue as the Glisson's capsule over the surface of the liver and parietal peritoneum on the abdominal cavity. These ligaments are usually thin and avascular in the healthy state. Dividing these ligaments is the initial step in any major liver surgery or liver transplantation.

1.2 Vascular and Biliary Anatomy

The vascular anatomy of the liver in children is every similar to that in adults.

- The common hepatic artery arises as a branch of the coeliac artery and proceeds to the liver after giving the gastroduodenal artery.
- The portal vein is formed by the confluence of the superior mesenteric vein and the splenic vein.
- Both these structures enter the liver at the hilum after passing through the left edge of the gastrohepatic ligament.

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