Ain Shams University
Faculty of Medicine
Department of Anesthesia, Intensive care and Pain Management

Anesthetic Considerations for Pediatric Liver Transplantation

Essay

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By

Salma Ahmad Abd Elkader Sheesh
M.B. B CH. Ain Shams University

Supervised by

Dr. Gamal Eldin Mohammad Ahmad Elewa
Professor of Anesthesia and intensive care
Faculty of Medicine – Ain shams University

Dr. Ashraf Elsayed Abd ElRahman ElAgamy
Lecturer of Anesthesia and intensive care
Faculty of Medicine – Ain shams University

Dr. Ayman Ahmed El-Sayed Kasem
Lecturer of Anesthesia and intensive care
Faculty of Medicine – Ain shams University

Faculty of Medicine

Ain shams University
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فى حالات زراعة الكبد للأطفال

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الطبيبة : سلمي أحمد عبد القادر شيش
بكالوريوس الطب والجراحة العامة – جامعة عين شمس

تحت إشراف
الأستاذ الدكتور / جمال الدين محمد أحمد عليوه
أستاذ التخدير والرعاية المركزة
كلية الطب- جامعة عين شمس

الدكتور / أشرف السيد عبد الرحمن العجمي
مدرس التخدير والرعاية المركزة
كلية الطب- جامعة عين شمس

الدكتور / أيمن أحمد السيد قاسم
مدرس التخدير والرعاية المركزة
كلية الطب- جامعة عين شمس

كلية الطب

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Aim of The Work

The aim of this work is to throw light on the most common causes of liver transplant in pediatric population and how to do proper preoperative assessment and preparation for surgery , and proper intra-and post-operative management for such patients .

List of Abbreviations

α-IATD	Alpha 1 antitrypsin deficiency
aPTT	Activated partial thromboplastine time
ATP	Adenosine triphosphate
BUN	Blood urea nitrogen
CCI	Color coded imaging
CF	Cystic fibrosis
CMV	Cyto megalo virus
CNIs	
CPM	Central pontine myelinolysis
CVP	Central venous pressure
CSA	Cyclosporine A
DNA	Deoxyribo nucleic acid
EACA	E-aminocaproic acid
EBS	Epstein Barr virus
ECG	Elecrto cardio gram
ESLD	End stage liver disease
FFP	Fresh frozen plasma
FHF	Fulminant hepatic failure
FNH	Focal nodular hyperplasia
FRC	Functional residual capacity

GSD	Glycogen storage disease
HAV	Hepatitis A virus
HAT	Hepatic artery thrombosis
HB	Hepato blastoma
HBV	Hepatitis B virus
HCC	Hepato cellular carcinoma
HCV	Hepatitis C virus
HDV	Hepatitis D virus
HIDA	Hepatobiliary imino-Diacetic acid
HPS	Hepato pulmonary syndrome
ICP	Intra cranial pressure
IVC	Inferior vena cava
i.v.	Intra venous
LT	Liver transplantation
MA	Maximum amplitude
MELD	Model for end stage liver disease
NH	Neonatal hemochromatosis
NIDDK	National institute of diabetes and digestive and kidney diseases
NTBC	Nitro trifluoromethyl benzyl cyclohexendione
OLT	Orthotopic liver transplant
OPTN	Organ procurement and transplantation Network
OR	Operating room

PAP	Pulmonary artery pressure
PCR	Polymerase chain reaction
PEEP	Positive end expiratory pressure
PELD	Pediatric end stage liver disease
PICU	Pediatric intensive care unit
PRBCs	Packed red blood cells
PT	Prothrombim time
PTT	Partial thromboplastin time
RNA	Ribo nucleic acid
RIS	Rapid infusion system
PNF	Primary nonfunction
SD	Standard deviation
SFHF	Sub acute fulminant hepatic failure
SVR	Systemic vascular resistance
TBA	Tissue plasminogen activator
TEE	Trans esophageal echo cardiography
TEG	Thrombo elastogram
TNF α	Tumor necrosis factor alpha
TTI	Tyrosinemia type 1
UNOS	United network for organ sharing
VVB	Veno venous bypass
WD	Wilson disease

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INTRODUCTION

End-stage liver disease in pediatric may be due to congenital causes (e.g. biliary atresia) or acquired disease (e.g. hepatic tumors) (Hammer and Krane , 2001). Since the early 1990s liver transplantation has established itself as a highly successful treatment for the management of end-stage liver disease (Biggins et al ,2005).

Cases of Pediatric liver transplantation are becoming increasing with time. Recent advances in the surgical and anesthetic management of these cases have greatly improved the outcome of these cases. In order to successfully manage the anesthesia in these patients, one needs to have a thorough understanding of the pathophysiology of end-stage liver disease and the subsequent anesthetic implications. It is also necessary to appreciate the stages of the surgical procedure, as each stage presents different dilemmas to the anesthesiologist. Thus, anethesiologist should review the pathophysiology of liver failure in pediatric patients and outline the particular issues related to each stage of liver transplantation, allowing for the anticipation and management of the derangement that occurs during surgery (Yudkowitz and Chietro , 2005).

The care of a child undergoing liver transplantation poses one of the greatest challenges in modern medicine (Biggins et al , 2005).

The first successful pediatric liver transplant was performed in 1967. Between 1989 and 2004, almost 1000 pediatric patients have successfully undergone liver, pancreases, or intestinal transplantation in the United States. Before 1980, with the use of the immunosuppressive agents azathioprine and prednisone, the 5-years survival in the pediatric patient following liver transplantation was a dismal 20% (Gordon, et al, 1991).

After the introduction of cyclosporine in 1980, long-term survival after liver transplantation become a reality. For the first time, 5-year patient survival began to exceed that of the life expectancy related to the specific disease process. Graft survival has progressively improved since 1992. For example, a 1-year graft survival was 81% in 2001 (6-to 10-year recipient age) compared with 68% a decade ago. Patient survival for this age group is estimated at 1 year to be 90.5%; 3 years, 85.9% ; and 5 years , 83.8%. Patient survival in the first year after transplant is similar for all age groups except children younger than 1 year, who have the highest annual death rate. For these infant recipients transplanted in 2001 and 2002, there was a marked decline in 1-year death rate, which is also seen as a trend for children aged 1 to 5 years and those aged 11 to 17 years. Improvements in patients and graft survival rates have been attributed to new immunosuppressive regimens (UNOS/OPTN, 2005).

INDICATIONS FOR PEDIATRIC LIVER TRANSPLANTATION

Liver disease in childhood produces a variety of symptoms affecting a range of children of all sizes, from preterm neonates to adolescents. Thus, there are conditions specific to the neonatal period such as neonatal hemochromatosis, and conditions affecting infants, for example, extrahepatic biliary atresia. With increasing age, a more heterogeneous disease pattern emerges with conditions such as α-1 antirypsin deficiency and autoimmune hepatitis becoming prevalent as well as diseases that are also found in the adult. In addition, the condition may present acutely or in a chronic manner (Dhawan, et al, 2004).

Indications for liver transplantation (LT) in children include progressive subacute or chronic primary liver disease, such as biliary atresia; metabolic diseases of the liver; fulminant hepatic failure, hepatic tumors, and retransplantation for hepatic graft failure (Table 1) (Marcel, et al, 1996).

The most common disorder for which LT is performed in children is biliary atresia, accounting for more than 50% of patients . It is the most common cause of chronic cholestasis in infants and children, affecting an estimated 1 in 8,000-12,000 live births worldwide (Rykman, et al, 1997).

Table 1: Indications for pediatric liver transplantation

Indication	*Examples (%)*
Subacute / chronic liver disease	Biliary atresia/hypoplasia (46%) Cirrhosis (postnecrotic, cryptogenic) (9%) Cirrhosis 2° to TPN (3%) Neonatal hepatitis (3%)
Metabolic liver diseases	1. Alpha-l-antitrypsin deficiency (5%) 2. Tyrosinaemia (1.5%) 3. Wilson's disease (1.3%) 4. Glycogen storage disease (0.5%) 5. Haemochromatosis (0.5%) 6. Extrahepatic presentation 7. Oxalosis (1%) 8. Protein C, S, antithrombin III deficiency (< 0.5%)
Fulminant hepatic failure	Viral hepatitis (2%) Drugs, toxins (1.2%)
Hepatic tumors	Malignant (2%) Hepatocellular carcinoma Hepatoblastoma Benign (0.7%) Haemangioendothelioma
Hepatic graft failure	Retransplantation (1 %)

(Marcel et al, 1996)