INTRODUCTION

• The treatment of end stage liver disease underwent a dramatic transformation with the development of liver transplantation.

• Operative procedures that were experimental and treacherous 2 decades ago are now being performed successfully and are considered the standard of care for patients with liver failure.

• Pediatric liver transplant is one of the most successful solid organ transplant with UNOS reporting 1 and 3 years patient survival approaching and exceeding 90% and 80% respectively.

• However, the risks of the operation remain considerable, so careful consideration must be given to the indications and timing of the procedure, and meticulous preparation of the recipient is necessary.

• In this talk, I will review the stages of evaluation of the candidate beginning with the:
  – Goals of evaluation
  – Indications for and contraindications to LT
  – Medical evaluation of the candidate
  – Process of listing
Goals of Evaluation

- The primary aims of the evaluation process are to define which patients require or would benefit from LT and when such therapy should be undertaken.
- It is important to ensure that other relevant medical and surgical treatments have been considered and that LT remains overall the best option.

General Indications for LT in Children

- Biliary atresia is by far the most frequent single indication for LT.
- Chronic hepatitis, certain metabolic diseases, acute liver failure are also common indications.
- Cirrhosis is an indication for immediate LT when there is evidence of liver decompensation such as:
  - Abnormal blood clotting
  - Fluid within the abdominal cavity
  - Frequent or massive intestinal bleeding
  - Malnutrition or growth failure
  - Frequent bacterial infections

- We usually direct the evaluation towards a precise understanding of the underlying liver disease:
  - Identification of progressive deterioration in liver function
  - The development of complications such as portal hypertension, recurrent infection or gastrointestinal bleeding
  - Nutritional/growth failure

Other goals are optimization of nutritional and medical therapy for the child awaiting LT, providing education to the family and finally predicting the optimal timing of LT.

- Patients known to have cirrhosis should be followed at regular intervals to monitor growth, progression of their disease and to determine the optimal time for LT.
Contraindications to Pediatric LT

• As surgical techniques and medical management have improved, the list of contraindications has shortened.
• Absolute contraindications:
  – Uncontrolled infection
  – AIDS
  – Irreversible massive brain injury
  – Tumor outside the liver that cannot be removed
  – Tumor from another organ that has spread to the liver

Relative contraindications:
  – Partially treated infection
  – HIV infection

Medical evaluation of the candidate

• Begins with recognition of the patients original diagnosis and assessment of complications arising from liver failure, enabling the team to determine the need for and the urgency of LT.

Blood work:
  – hematology (blood count, clotting study, blood group and antibody screen);
  – chemistry (liver tests, vitamin A,D,E levels);
  – serologies (Hepatitis A,B,C, CMV, EBV, varicella, measles, HIV)

Radiological studies:
  – ultrasound of the blood vessels of the liver

Consultation with specialists:
  – transplant surgeon
  – transplant coordinator
  – nutritionist
  – anesthesiology
  – social worker
  – psychologist
  – medical consults as needed
• The evaluation addresses psychological factors in terms of the child’s understanding of his/her illness and the parents understanding of the transplant process and their child’s prognosis.

• The concept of living related donation should be introduced during the evaluation process.

• This is an important alternative for families with a child facing LT.

• The advantage to this procedure is two-fold:
  – Quality of the graft
  – Opportunity to schedule the transplant while the patient is still an optimal candidate

• The donor must be a healthy adult <50-55 years of age, preferably a parent, and have a compatible blood type.

• The donor must undergo a complete medical evaluation, must be judged to be emotionally stable and have a reliable support network of family and friends who will be available during the surgical procedure and until the donor is fully recuperated from surgery.

• Every attempt should be made to include the child in age-appropriate discussion of LT during the evaluation process.

• Once the need for LT is established and no contraindications exist, patients are listed for deceased donor regardless of whether a potential living donor exists.

• All patients accepted onto a transplant’s hospital’s waiting list are registered with the united network of organ sharing (UNOS).

• UNOS is staffed 24 hours a day throughout the year and assists with the matching, transporting, and sharing of organs throughout the United States.

• Prior to 2002, priority for deceased donor liver allocation in the UNOS system was primarily based on waiting time.
• In Feb. 2002, UNOS adopted the MELD/PELD allocation system, which was designed to prioritize patients by acuity of illness rather than waiting time.

• In this system, PELD and MELD scores rank children and adults on a single waiting list according to their probability of death within 3 months of listing.

• Both scores are based on the same underlying principles and rely on objective and measurable parameters.

Management of the child awaiting LT

• Once a child has been placed on the waiting list for LT, it remains important to optimize all aspects of the patient's care prior to surgery.
• Management of complications of liver disease should continue as per standard of care.
• Particular attention should be placed on enhancing the child's nutrition.
• General pediatric follow-up can also help in general development, hearing and vision screening and infection surveillance.
• Complete immunization including all standard immunization is of utmost importance.

Referral to a transplant center

• The best time for referral is as soon as the patient is identified as having a condition that will require transplantation e.g.
  – Patients with biliary atresia who remain jaundiced after a Kasai procedure
  – Metabolic disease poorly controlled with medication
  – Cirrhosis from any reason.

Early referral allows the transplant center to have maximum input into the pre-transplant management strategy.

• When donor organs are identified, the procuring organization accesses the UNOS computerized organ matching system, enters information about the donor organs and the computer generates a list of potential candidates.
• The procurement coordinator contacts the transplant surgeon caring for the top ranked patient to offer the organ.
• Depending on various factors such as the donor's medical history and the current health of the potential candidate, the transplant surgeon determines if the organ is suitable for the patient.
• Once the organ is accepted, the candidate is identified prior to organ recovery and called into the hospital where the transplant will occur to prepare for the surgery.

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• Both scores are based on the same underlying principles and rely on objective and measurable parameters.
• The mutual goal is to develop a close working relationship between the family, the pediatrician and the transplant center as this type of partnership is essential for the optimal coordination of post-op care.

Summary

• Evaluation of the candidate for pediatric LT is a vital step in ensuring a successful outcome from surgery.
• Requires a multidisciplinary approach and involves an assessment of the candidates underlying illness and indication for LT, the severity of liver disease, the timing of LT, and a medical and psychosocial evaluation.
• Continued careful evaluation of candidates combined with a listing mechanism that allocates organs to the sickest individuals first will ensure that pediatric LT remains one of the most successful solid organ transplants.