Liver transplantation in children

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https://ekids.hcuge.ch/
Outline

• Commonly accepted indications
• Surgical aspects
• Allocation
• Outcomes
Demographics of pediatric liver transplantations in Switzerland (1989 – 2014)

- **Number of patients**: 140 (8 Re-LTx)
- **Number of LTx**: 137
- **Age at LTx**: 3.3 months - 16.8 years: median: 21 months
- **Weight**: 3.5 - 59 kg (median: 10.3 kg)
  - 1/2 < 10 kg
  - 2/3 < 15 kg
Age at liver transplantation (Bicêtre, Paris)

REF: D. Debray, Paris

n = 648 LTx
Indications for pediatric liver transplantation

- Fulminante
- AVB (Atrésie des voies biliaires)
- PFIC
- Métabolique
- Choléstase
- Divers
- Re-tx

Indications at Pittsburgh: 808 patients; Transplantation, 2002, 73(6)
Liver transplantation for biliary atresia

• Most common indication in childhood
• Life saving (Starzl)
• No recurrence of disease on graft
• 2 indications
  ▪ Liver failure in infancy
  ▪ Portal hypertension in children & adults
LT for liver-based metabolic disease

- 2\textsuperscript{nd} most common indication in children
- Basic principle: gene therapy
- Outcome depends on gene expression
LT for liver tumors

**Hepatoblastoma**
- Pretext IV
- Transformed survival:
  - 100% 1 year
  - 83% 4 years
- Few contraindications
  - Lung metastases
  - Vascular invasion

**Hepatocellular carcinoma**
- Rare in children
  - Except Tyrosinemia type I
- Outcome ≠ adults
- Incidental, in situ
- Milan criteria do not apply

Héry et al 2011
Otte 2010
Romano 2011
Hadzic 2011
Ismail 2009
Sevmis 2008
LT for quality of life

• Pruritus
  ▪ Alagille syndrome
  ▪ Genetic cholestasis

• Liver-based metabolic diseases
  ▪ Gastrostomy feeding
  ▪ Special diets
Contra-indications for LT

• Irreversible, progressive extrahepatic disease
  – Mitochondriopathies?
  – Niemann Pick C

• Non-controlled malignancy

• High risk of recurrence on graft?

• Cryptogenic disease (acute or chronic)

• Mental retardation?
LT in children with developmental delay

- **Survival**
  - 100% 1 year
  - 90% 3 years

- **Good compliance**
  - Caregivers

Martens et al Pediatr Transplant 2006,
Korsch 2006
The graft in pediatric liver transplantation

Whole liver

Shared liver = split
The graft

- Whole liver: 2% body weight
- Left lobe: 25% - 30% of total liver weight
Graft types: Adults vs. children (Geneva 1989 - 2013)

Adults (n=556)

- Foies entiers: 91%
- Foies partagés (>1999): 4%
- Foies réduits: 4%
- Foie donneurs vivants: 1%
Graft types: Adults vs. children
(Geneva 1989 - 2013)

Adults (n=556)
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- Foie donneurs vivants: 1%

Children (n=137)
- Foies entiers: 35%
- Foies partagés (>1999): 24%
- Foies réduits: 33%
- Foie donneurs vivants: 8%
Organ allocation

**Position on waitlist**

- MELD (creatinine, INR, bilirubin)

**Priority**

- „normal“
- „super-urgent“:
  - Acute liver failure
  - Hepatoblastoma
  - Re-Tx for primary non-function
**Allocation in Switzerland (2013)**

- Super urgent status
- Donor < 50 y/o $\rightarrow$ recipient < 25kg
- Donor < 18 yrs (Art. 11)
  - Child < 12 years
  - Child 12-18 years
  - Recipient > 18 years with highest MELD

No death on waiting list since 2007 (SOAS)
Acceptance criteria in children

• ABO
• No transmissible disease
• **Donor Age**: <50 years greater than child
• Organ quality:
  – History
  – Lab values
  – Ultrasound
  – **Liver volume**
Graft volume

• Weight donor/recipient
  - ≤ 2  => whole liver
  - 2 - 4  => left liver (segments II/III/IV)
  - 4 - 10  => left lobe (segments II/III)
  - ≥ 10  => monosegment
  - for exemple 60 kg / 6 kg = factor 10

• Weight donor liver / recipient
  - Normal: 5% (newborn) - 2% (adult)
  - Recommended: 1% - 3%
  - for example 300 g / 6 kg = 5%
Implanted left lobe in a child with biliary atresia
Survival following LT in Geneva

Patient survival

Graft survival

= 90%

= 82%
Survival following LT
Split vs. whole graft

Patient survival

Graft survival

Split liver
Whole liver
p = 0.629

Split liver
Whole liver
p = 0.707
It is not only about survival...

...we got to think about growth, development and quality of live after LT!

Source: B. Otte, Bruxelles
Cognitive function and psychosocial adaptation

• **Cognitive function**
  - Many children are not at the same level of functioning as their IQ would predict.
    - ⇒ *Learning difficulties*, problems at school

• **Psychosocial adaptation**
  - **Quality of life** after pediatric LT is comparable with that of children with *chronic disease*
  - Yet, it is worse than that of healthy children

Adèle, 10 years after pediatric liver transplantation
Conclusion

- Liver transplantation is **lifesaving** for children
- **Unique indications** in pediatrics
  - ↓ risk of recurrence on the graft
- Children well served by **Swiss allocation system**
- Advantages of **split liver**
- **Excellent** long term **survival**
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