Long term home parenteral nutrition ... or small bowel transplantation

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Intestinal failure: definitive

- Ultra-short bowel:
  ± < 30 cm - valve
  ± < 20 cm + valve
- Microvillous atrophy
- Long segment Hirschsprung disease
  (= total aganglionosis)
- Tufting enteropathy?
Intestinal failure: not definitive?

• Short bowel
  – 40 -> 80 cm: adaptation; lengthening?

• Tufting enteropathy
  – a few weaned from PN

• CIPOS
  – nursing, surgery; prognosis?
First goal of care = feed the child

Parenteral nutrition

– sustain growth and development
– promote intestinal adaptation
– no additional complications

• Bridge to adaptation and normal function
• Long term treatment
What is the future?

- Long term parenteral nutrition
  - transplantation only if complications

- PN as a bridge for small bowel transplantation
Survival on home PN / indication

### Prognosis: Necker 1980-2000, n=302

<table>
<thead>
<tr>
<th></th>
<th>SBS</th>
<th>Diarrhea</th>
<th>CIPOS</th>
<th>IBD</th>
<th>Imm. def</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Weaned</strong></td>
<td>61</td>
<td>12</td>
<td>32</td>
<td>85</td>
<td>46</td>
</tr>
<tr>
<td><strong>Death</strong></td>
<td>7</td>
<td>24</td>
<td>13</td>
<td>3</td>
<td>39</td>
</tr>
<tr>
<td><strong>Transplant</strong></td>
<td>6</td>
<td>16</td>
<td>3</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td><strong>Still on PN</strong></td>
<td>26</td>
<td>48</td>
<td>52</td>
<td>12</td>
<td>15</td>
</tr>
</tbody>
</table>
SBS: weaning off PN 2000-2010

• Median: 19 months

• Negative factors
  • Univariate
    o length of the remnant bowel < 40 cm
    o absence of ICV, colon < 50%
    o IV Lipid > 1.5g/kg/d, PN caloric > 70%

• Multivariate: length of remnant bowel < 40 cm
Complications of home PN

- Home PN is impossible
- Extensive vascular thrombosis
- Multiple infections
- Progressive IF-associated liver disease
- Growth failure
- Psychological intolerance / HPN
Complications of home PN

- *Home PN is impossible*
- Extensive vascular thrombosis
- Multiple infections
- Progressive IF-associated liver disease
- Growth failure
- Psychological intolerance / HPN
Home PN impossible?

- The aim of our meeting!!
- Social and psychological help
- Limits:
  - long term hospitalization
  - microvillous atrophy: unstability ++

= early indication
Complications of home PN

- Home PN is impossible
- Extensive vascular thrombosis
- Multiple infections
- Progressive IF-associated liver disease
- Growth failure
- Psychological intolerance / HPN
Vascular thrombosis?

• Controlled size and position of central lines

• Limits:
  – elsewhere …
  – 2 thrombosis
  – Do not wait for the last central line
superior and inferior cave thrombosis

Just one left subclavian vein

Transliver KT

Under-renal thrombosis
Complications of home PN

- Home PN is impossible
- Extensive vascular thrombosis
- *Multiple infections*
- Progressive IF-associated liver disease
- Growth failure
- Psychological intolerance / HPN
Infections?

- Staff and caregiver training
- Taurolidine / ethanol locks

- Limits:
  - diarrhea ++
  - child’s behaviour
  - hospitalization elsewhere …
Complications of home PN

- Home PN is impossible
- Extensive vascular thrombosis
- Multiple infections
- Progressive IF-associated liver disease
- Growth failure
- Psychological intolerance / HPN
Progressive liver disease?

- Control of risk factors
  - + fish oil
- Limits:
  - elsewhere …
  - sometimes it does not work …

- Necker: +/- 110 children home PN (40% France)
  - 2 end-stage liver diseases in last 10 years
Mortality on long term PN

= 16 %

  9 % : digestive disease
  38 % : other (AIDS …)

Cause

  – original disease : 62 %
  – infection : 11 %
  – liver : 12 %
  – other : 15 %

Necker-EM 302 patients
IF-associated liver disease is reversible (without fish oil)

- Liver 1999

99-2002: home PN

- Liver 2002
Complications of home PN

- Home PN is impossible
- Extensive vascular thrombosis
- Multiple infections
- Progressive IF-associated liver disease
- Growth failure
- Psychological intolerance / HPN
Growth?

- Adapted nutrients and energy

- Limits:
  - liver?
Complications of home PN

• Home PN is impossible
• Extensive vascular thrombosis
• Multiple infections
• Progressive IF-associated liver disease
• Growth failure
• Psychological intolerance / HPN
Psychological intolerance?

- Discuss risks!
- Psychological help

But:
- you do not understand before being in …
- intolerance may => complications
  - less care?
Home PN *versus* Tx

- HPN ———

- HPN ——— //

  Tx ———— .... >
Small bowel Tx: indications

Total AND definitive intestinal failure

AND

complications of home PN

Do not wait for major complications!

!! Waiting time
Indications

Children

- Short Gut: 63%
- Gastrochisis: 21%
- Trauma: 1%
- N. Enterocolitis: 12%
- Volvulus: 16%
- Ischemia: 1%
- Atresia: 4%
- Other Short Gut: 3%
- Other: 5%
- Retransplant: 8%
- Malabsorption: 9%
- Tumor: 1%
- Motility Disorder: 19%
- Other: 1%

Intestinal Transplant Registry Report © 2015

- 9 children
- 102 children, 111 transplantations
  - short bowel : 35
  - congenital enteropathy : 35
  - motility disorder : 28
  - re-transplantation : 10

- 102 children, 111 transplantations
- Age at Tx : +/- 4.5 y
  - 59 : isolated small bowel Tx
  - 44 : combined-liver small bowel
  - 4 multivisceral Tx, 1 modified multivisceral Tx
  - + colon : 67
Which type of transplantation?

Isolated small bowel Tx vs combined liver-SB Tx

- more organs
- less early mortality

- less early rejection
- less late graft losses

$\Rightarrow$ liver protects intestine?
Do a liver biopsy

- Normal --> F2
  - Isolated small bowel
  - Control risk factors +++
  - Biopsy / 6-12 months

- F3-4
  - + liver
Type of transplantation

- Isolated small bowel Tx
  - short bowel syndrome
- Small bowel + colon
  - congenital enteropathy
  - motility disorder
- Small bowel + colon + stomach (+ pancreas)
  - CIPOS, very long-segment Hirschsprung
Type of transplantation

• Liver + small bowel Tx
  • idem small bowel + severe fibrosis

• Multivisceral
  • motility disorder + severe fibrosis
Difficult discussions …

Isolated liver transplantation

– ESLD + SBS + normal morphology

– realistic expectation of gut adaptation
  o previous enteral tolerance : 50 %
  o age < 4 y
Small bowel transplantation

- Oldest survivor with graft:
  - French, born 1988, Tx 1989
- Most patients: < 15 y F/U
- Risks and complications
  - many !!!
- Long-term: % late graft loss?
Graft Survival by Era

**Children**

- 2000-2005
- 2000-2005
- 1985-1999

**Adults**

- 2006-2014
- 2000-2005

p=<0.001
Patient survival by transplant type: children (2009-2014)
Graft survival by transplant type: children (2009-2014)
Functional Status
(Patients Transplanted 2009 – 2014 who have survived at least 6 months)

**Freedom from PN**

**Modified Karnofsky Performance Scores**

- 40 patients with graft
  - off PN, +/- enteral nutrition

- 17 returned to PN

- 5 patients on the waiting list
In conclusion

• 2016: long term PN best prognosis
  – if PN in intestinal failure centre
• If you must transplant:
  – do not wait for major complications
  – will be EVEN MORE difficult

• More children die early after L-SBTx
• More grafts survive late after L-SBTx
Teşekkür ederim!

10 years post-L+SBT