Pediatric Intestinal Transplantation Questionnaire
Dear colleague,

This questionnaire will mainly focus on the referral criteria for pediatric intestinal transplantation in children (0-18 years). It evaluates the considerations when children are referred for intestinal transplantation.

It consists of three parts:

Part I  General inquiries  (consists of 18 questions)
Part II  Referral criteria  (consists of 16 questions)
Part III  Cases     (consists of 3 cases)

It will take about 30 min to fill in this questionnaire.

All information will be dealt with anonymously and discretely.

There are three possibilities to fill in this questionnaire:

- Fill in online: a link has been sent to your email address. If you would like to receive your link again, please send an email to the email address below.

- Fill in this paper version of the questionnaire and return it in the included envelope, NO stamp is required. If you have not received a paper version, and if you prefer the paper version, please send an email to the email address below.

This questionnaire is part of a Masters’ thesis for the MSc in Surgical Sciences at the University of Edinburgh. We would greatly appreciate it, since this thesis is restricted to time limits, if you could complete this questionnaire by: April 8, 2011.

In case you have any questions, please contact Marie-Chantal Struijs, either by email: a.e.c.j.m.struijs@erasmusmc.nl or phone: +31610897076.

Thank you very much in advance for your cooperation!

With kind regards,

Prof JNM IJzermans, MD, PhD    Marie-Chantal Struijs, MD    Dr VB Nieuwenhuijs, MD, PhD
Transplant Surgeon              Research fellow              Transplant Surgeon
Director of Surgical Training   Erasmus MC, Rotterdam     UMC Groningen
Inquiries about the person filling in this questionnaire:

I am a   ☐ Gastroenterologist
        ☐ Surgeon
        ☐ Nurse practitioner
        ☐ Other, __________

I work at a   ☐ Pediatric transplant center
             ☐ Adult transplant center
             ☐ Both

This center is situated in:   ☐ USA, Canada
                                ☐ Europe

I work at different pediatric transplant centers:   ☐ No
                                                      ☐ Yes

My age is: __________years

My gender is  ☐ Male
              ☐ Female

I have ________years of experience in this field

Please start the questionnaire
PART I

GENERAL INQUIRIES
Question 1

In which year was the first pediatric **intestinal** transplantation (0-18 years) performed in your center?

- [ ] ___________ → continue to question 3
- [ ] NA → continue to question 2

Question 2

This question is only applicable if you do **not** perform pediatric **intestinal** transplants.

What is the reason no intestinal transplants are performed?

*(multiple options are possible)*

- [ ] Marketing
- [ ] Patient population too small
- [ ] Procedure too difficult
- [ ] Not a program priority
- [ ] Financial
- [ ] Other ____________________________

______________________________

______________________________

If you do not perform pediatric intestinal transplants, please continue to Part II Referral criteria
Question 3
Which kind of pediatric visceral transplants are performed in your center?
(multiple options are possible)
- Isolated intestinal transplantation
- Isolated liver transplantation
- Combined intestinal-liver transplantation
- Multivisceral transplantation (any combination)

Question 4
Regarding question 3, please indicate the number of transplants which were performed in your center in patients under 18 years in the last 5 years (2006-2010)

______________ number of isolated intestinal transplants
______________ number of isolated liver transplants
______________ number of combined intestinal-liver transplants
______________ number of multivisceral transplants

Question 5
How many surgeons are performing pediatric visceral transplants in your center?
______________ surgeons

Question 6
The majority of patients are referred to our center for:
- Assessment for transplant
- Comprehensive treatment of intestinal failure
- Both options equal number of patients
### Question 7

What are the **3 main reasons/diagnosis** patients are referred for transplant to your center (column 1) and what are the main diagnosis for patients undergoing intestinal transplantation at your center (column 2)?

*Please put 1, 2, and 3 behind the 3 main reasons, with 1 being the most common reason, 2 the 2nd most common and 3 for the 3rd most common reason*

<table>
<thead>
<tr>
<th>Column 1</th>
<th>Column 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Referral for assessment for intestinal transplant</td>
<td>Diagnosis at intestinal transplantation</td>
</tr>
<tr>
<td>Short bowel syndrome due to volvulus</td>
<td></td>
</tr>
<tr>
<td>Short bowel syndrome due to gastroschisis</td>
<td></td>
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<tr>
<td>Short bowel syndrome due to necrotizing enterocolitis</td>
<td></td>
</tr>
<tr>
<td>Short bowel syndrome due to atresia</td>
<td></td>
</tr>
<tr>
<td>Short bowel syndrome due to other reasons</td>
<td></td>
</tr>
<tr>
<td>Motility disorders (Hirschsprung, pseudo-obstruction)</td>
<td></td>
</tr>
<tr>
<td>Mucosal disorders (i.e. microvillus inclusion disease)</td>
<td></td>
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<tr>
<td>Retransplantation</td>
<td></td>
</tr>
<tr>
<td>Other:</td>
<td></td>
</tr>
</tbody>
</table>

### Question 8

With the improvement in medical (ie better forms of TPN) and surgical care (ie intestinal lengthening procedures) do you think there continues to be a **role** for intestinal transplantation?

- [ ] YES
- [ ] NO
Question  9

Regarding question 8, do you think the role for intestinal transplantation will:

- Increase over time
- Decrease over time
- Remain the same

Question  10

Also, with the improvement in medical and surgical care, do you notice a difference in the number of patients referred for intestinal transplantation?

- Increased
- Decreased
- Remains the same

If you have answered “remains the same” skip question 11

Question  11

What do you think is the reason the number of patients referred for intestinal transplantation has changed (either increased or decreased)?

- Improvement in medical care (i.e. TPN)
- Improvement in surgical procedures (i.e. intestinal lengthening procedures)
- Both
- Other, ________________________________  
  ________________________________  
  ________________________________

Question  12

How many patients have been referred to you the last 5 years (2006-2010) for intestinal transplantation?

± ___________________________ patients
SUPPLEMENTAL DIGITAL CONTENT 1

Question 13

What do you think of this statement:

I believe patients should be referred for intestinal transplant at an earlier time than patients are referred at this moment

☑ Strongly disagree
☑ Disagree
☑ Neither agree or disagree
☑ Agree
☑ Strongly agree

Question 14

What percentage of patients that are referred are not suitable for intestinal transplantation for medical and/or surgical reasons?

______________%

Question 15

What do you think the reason is that these patients are not suitable?

☑ Incorrect use of the referral criteria
☑ Bad communication between referral hospital and transplant center
☑ Referring doctors are not aware of indications for referrals
☑ Referring center wanted a comprehensive assessment of intestinal failure treatment options, not just a transplant
☑ Other, __________________________

_____________________________________

_____________________________________

Question 16

What percentage of patients that are referred to your center, should have visited your center earlier in your opinion?

______________%
Question 17
Do you believe intestinal lengthening procedures should be performed at the referral center, if possible, before the patient is referred for intestinal transplantation?

☐ YES
☐ NO

Please explain your answer: ______________________________________________________
____________________________________________________________________________
____________________________________________________________________________

Question 18
Do you believe intestinal lengthening procedures should solely be performed in transplant centers?

☐ YES, only in transplant centers
☐ NO, also possible in the center that refer the patient
☐ NO, only in high-volume centers

Please explain your answer: ______________________________________________________
____________________________________________________________________________
____________________________________________________________________________
PART II

REFERRAL CRITERIA
Question 1
Is there a document available in your center which lists the criteria for referral/listing for intestinal transplantation in children (0-18 years)?

☐ YES (if yes, please attach this document to this questionnaire, please continue to question 2)

☐ NO (please continue to question 5)

Question 2
Is this document available online (on the internet)?

☐ YES

☐ NO

If no, please explain why this is not available online: ________________________________

__________________________________________

__________________________________________

Question 3
Did you sent this document to hospitals in your region which refer patients to your hospital?

☐ YES

☐ NO

Question 4
Are these criteria adjusted to the wishes of the surgeons and gastroenterologists in your center or are these the same as the criteria from the literature/American Society of Transplantation?

☐ Criteria are adjusted to the preferences of the surgeons and gastroenterologists

☐ Criteria are taken from the literature/American Society of transplantation

☐ Other, ________________________________
Question 5 Referral Criteria

In the literature (Avitzur et al. 2010) the following criteria are listed as the recommended pediatric referral criteria for intestinal transplantation:

- Children with massive intestinal resection
- Children with severely diseased bowel and unacceptable morbidity
- Microvillous inclusion disease or intestinal epithelial dysplasia
- Persistent hyperbilirubinemia (>6 mg/dl or > 100 µmol/l)
- Thrombosis of 2 of 4 upper body central veins
- Continuing prognostic or diagnostic uncertainty
- Request of the patient and family

The following questions will consider these criteria.

5a

Are these the criteria that are used in your center?

☐ YES
☐ NO

If no, please elaborate on why these referral criteria are not used in your center: 

5b

What are the top 3 referral criteria that are used in patients referred to your center?

Please put 1, 2, 3 for the top three referral criteria

| Children with massive intestinal resection |   |
| Children with severely diseased bowel and unacceptable morbidity |   |
| Microvillous inclusion disease or intestinal epithelial dysplasia |   |
| Persistent hyperbilirubinemia (>6 mg/dl or > 100 µmol/l) |   |
| Thrombosis of 2 of 4 upper body central veins |   |
| Continuing prognostic or diagnostic uncertainty |   |
| Request of the patient and family |   |
5c
Do you think these criteria are sufficient/comprehensive?

☑ YES
☐ NO

Please explain your answer: ____________________________________________
_________________________________________________________________
_________________________________________________________________

5d
Do you believe these criteria are specific enough?

☑ YES, no changes are necessary
☑ NO, the criteria are too general: additions should be made to make them more specific

5e
Do you think all of these criteria should be present before the patients are referred?

☑ YES
☑ NO

5f
Do you think the referral should be adjusted to different age categories? For example, criteria for children <1 year, 1-6 years of age, and >6 years?

☑ YES
☑ NO

5g
Which criteria should definitely be present/are pivotal when the patient is referred for intestinal transplant to your center?

*Please put an X in the answer of your choice*

<table>
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</table>
5h

Please grade the criteria for importance, 1 being the most important criterion and 7 the least important criterion from the list (fill in 1, 2, 3, 4, 5, 6, and 7)

| Children with massive intestinal resection                        | 1  |
| Children with severely diseased bowel and unacceptable morbidity | 2  |
| Microvillous inclusion disease or intestinal epithelial dysplasia | 3  |
| Persistent hyperbilirubinemia (>6 mg/dl or > 100 µmol/l)          | 4  |
| Thrombosis of 2 of 4 upper body central veins                     | 5  |
| Continuing prognostic or diagnostic uncertainty                   | 6  |
| Request of the patient and family                                 | 7  |

5i

Which criteria should remain in the referral list and which ones should go out?

Please put an X in the answer of your choice and explain your answer

| 1. Children with massive intestinal resection                        | IN | OUT |
| 2. Children with severely diseased bowel and unacceptable morbidity |    |     |
| 3. Microvillous inclusion disease or intestinal epithelial dysplasia |    |     |
| 4. Persistent hyperbilirubinemia (>6 mg/dl or > 100 µmol/l)          |    |     |
5. Thrombosis of 2 of 4 upper body central veins | IN | OUT

6. Continuing prognostic or diagnostic uncertainty | IN | OUT

7. Request of the patient and family | IN | OUT

5j

Which criteria would you like to add to the referral list and why?

If you don’t have any criterion you would like to add, please fill in write on line 1

1

2

3

For example, prolongation of prothrombin time, recurrent septic episodes, severe fluid and electrolyte disturbances
Question 6

The following questions will consider the referral criterion:

*Persistent hyperbilirubinaemia (>6 mg/dl or > 100 µmol/l)*

6a

Do you think this is an important referral criterion for intestinal transplantation?

☐ YES
☐ NO

Please explain your answer: ____________________________________________

6b

Should this criterion be present in *every patient* referred for intestinal transplantation?

☐ YES
☐ NO

Please explain your answer: ____________________________________________

6c

The term “persistent” is not really defined. It is known from the literature that parenteral nutrition-induced liver disease leads to liver failure and death usually within 1 year. So should the referral criterion be changed to include time limit?

☐ YES
☐ NO

6d

Do you think changes should be made to the upper limit? For example change the >6mg/dl (> 100 µmol/l) into >4mg/dl (>67 µmol/l)?

☐ YES, it should be changed into: > _____ mg/dl or > _____ µmol/l
☐ NO
6e

It is known the prothrombin time becomes prolonged in the later stages of parenteral nutrition-associated liver disease. Do you think the prothrombin time should be included in the criteria?

☐ YES
☐ NO

6f

Do you think the criterion should be changed into: Intestinal failure-associated liver disease (IFALD), with conjugated bilirubin ≥2mg/dl (34.2 µmol/l)?

☐ YES
☐ NO

Question 7

The following questions will consider the referral criterion:

_Thrombosis of 2 of 4 upper body central veins._

7a

Do you think this is a pivotal referral criterion?

☐ YES
☐ NO

Please explain your answer: ____________________________________________________________
________________________________________________________________________________
________________________________________________________________________________

7b

One of the referral criteria is: Loss of more than 50% of the standard central venous access site; however one of the contraindications for transplant is insufficient vascular patency to guarantee vascular access.

Do you think this is contradictive?

☐ YES
☐ NO
Question 8

The following questions will consider the referral criterion:

Request of the patient and family

8a

Do you think this is an **pivotal** referral criterion in the decision-making process to refer a patient for intestinal transplantation?

- [ ] YES
- [ ] NO

Please explain your answer: __________________________________________________________

______________________________________________________________________________

8b

If the family would request an intestinal transplant but the patient does not fulfill all your criteria, would you **still** consider referral of the patient?

- [ ] YES
- [ ] NO

Please explain your answer: __________________________________________________________

______________________________________________________________________________

Question 9

*Microvillous inclusion disease or intestinal epithelial dysplasia*

These rare disorders almost never resolve or improve sufficiently with medical treatment. In most cases, death from liver disease or sepsis occurs within first 1-2 years of life.

Considering this information, do you think that having a congenital epithelial (mucosal) disorders without other criteria is sufficient for referral?

- [ ] YES, all patients should be referred as soon as the patient is diagnosed
- [ ] NO, more referral criteria should be present
Question 10

Small intestinal length < 30 cm with no ileocecal valve is mentioned as a risk factor for transplant.

10a

Do you think this should be incorporated into the referral criterion: Children with massive intestine resection?

☐ YES
☐ NO

10b

Do you agree with the following: children with less than 25% remaining intestine of the expected small intestinal length for age should be referred for transplantation when on TPN for more than 6 weeks?

☐ YES
☐ NO

10c

It is suggested the colon in continuity plays a role in intestinal adaptation. Moreover, enterocolonic discontinuity is suggested to be a risk factor for transplantation.

Do you think the following criterion should be added to the referral criteria: Enterocolonic discontinuity?

☐ YES
☐ NO

10d

Some articles define irreversible intestinal failure as to be dependent on TPN for >75% for more than 6 weeks and suggest to refer patients for transplantation.

Do you insist on referral of these patients as early as 6 weeks after being dependent on TPN?

☐ YES
☐ NO
SUPPLEMENTAL DIGITAL CONTENT 1

Question 11

Although recurring life-threatening sepsis are mentioned in the listing criteria, do you think physicians should insist on referral when children have recurring septic episodes (as was suggested in the article by Kaufman et al in 2001)?

☐ YES
☐ NO

Question 12

Refractory fluid and electrolyte disorders are mentioned as a risk factor for transplantation. Do you think this criterion should be added to the referral criteria?

☐ YES
☐ NO

Question 13

Outcome data show that mortality and morbidity associated with transplantation is higher when children are younger than 1 year of age. Do you think there should be a minimum age for referral for transplant?

☐ YES, it should be ____________ years
☐ NO

Question 14

The literature shows that children who are transplanted while still staying at home have a higher survival. Do you think children in general are referred too late for intestinal transplant?

☐ YES
☐ NO

What do you think should be done to change this (to achieve earlier referral)? (multiple options possible)

☐ Educate hospitals in your area
☐ More information on the internet
☐ Educate parents who have a child with intestinal failure
☐ Other, ____________________________________________
Question 15

Citrulline is identified as a potential biomarker of functional enterocyte mass. Do you think there should be a referral criterion considering biomarkers for intestinal function?

☐ YES
☐ NO

Question 16

Do you think it is useful to create a flow chart for the decision making process whether a patient should be referred for intestinal transplant?

☐ YES
☐ NO
PART III

CASES
CASE 1

Girl, 2\textsuperscript{nd} of monochorial-diamniotic gemelli, born at 37 weeks, birth weight 1350 grams. Six days after birth, midgut volvulus, 35cm ileal resection and jejunostomy. Six weeks later, ileocolic resection due to stenosis, and restoration of continuity using the remaining 45cm jejunum and colon. Other medical problems: hypothyreoidism requiring replacement therapy, muscle hypertonicity, developmentally delayed in motor skills, chronic anemia, and deficiencies of vitamin A and D, and zinc. Unable to discharge, in the hospital since birth. Parents also have the care for the patient’s twin sister and an older brother who is regularly admitted to the hospital. They are hesitant against prolonged admission in a transplantation center since they will not have the opportunity to visit often.

Current status: 2 ½ years old, weight 11.2 kg (0 SD) and height 72 cm (<-2.5 SD). Partially dependent on parenteral nutrition (50% enterally via drip feeding); Omegaven\textsuperscript{®} as lipid solution because of cholestasis. Thrombosis of one internal jugular vein; right and left subclavian vein have an “irregular” aspect. Multiple septic episodes (18x) and placement of 13 central venous catheters. Medication: questran, fraxiparine, vitamin supplements, imodium, thyrax, ranitidine. Last laboratory results: total bilirubin 5 µmol/l (0.3 mg/dl), direct bilirubin <1 µmol/l (<0.05 mg/dl), alkaline phosphatase 754 U/l, \( \gamma \)GT 76 U/l, AST 64 U/l, ALT 147 U/l, triglycerides 1.00 mmol/l.

QUESTIONS CASE 1

1
Is there medical information missing you need to evaluate this case for transplant?

- [ ] NO
- [ ] YES, namely ________________________________ ________________________________

2
Would you refer this patient for assessment for intestinal transplantation?

- [ ] YES (please continue to question 3)
- [ ] NO (please continue to question 4)
What are the reasons you would refer this patient?

Please put an X in the box of the referral criteria applicable to this patient

| Children with massive intestinal resection |  |
| Children with severely diseased bowel and unacceptable morbidity |  |
| Microvillous inclusion disease or intestinal epithelial dysplasia |  |
| Persistent hyperbilirubinemia (>6 mg/dl or > 100 µmol/l) |  |
| Thrombosis of 2 of 4 upper body central veins |  |
| Continuing prognostic or diagnostic uncertainty |  |
| Request of the patient and family |  |

Other reasons: ______________________________________________________

____________________________________________________

Please continue to question 5

What are the reasons you would not refer this patient?

- Not enough criteria to refer for transplant
- Still enough options for treatment
- Other, ___________________________________________________________________

____________________________________________________

What would be your treatment proposal?

- Medical treatment: try to increase enteral feeding
- Intestinal lengthening procedure
- Placement on waiting list for intestinal transplantation
- Other, ___________________________________________________________________

____________________________________________________
6

In case this patient would be medically suitable for intestinal transplantation, would the social situation (concerns of the parents) be of influence on your decision?

☑ YES, I would not place her on the waiting list
☑ NO, not of influence at all

7

Would you list this patient for intestinal transplantation?

☑ NO
☑ YES, based on the following criteria (please put an X in the boxes):

| Criteria                                                                 | 
|-------------------------------------------------------------------------|---|
| Small bowel length of <25 cm without an ileocecal valve                |
| Intestinal failure with high morbidity and poor quality of life        |
| Congenital intractable mucosal disorder                                |
| Persistent hyperbilirubinemia and signs portal HT or liver dysfunction |
| Loss of >50% of standard central venous access sites                   |
| Recurrent life-threatening episodes of sepsis                           |
CASE 2

Boy, born at term, no problems until 6 weeks after birth. Patient presented with intestinal cramps, vomiting and no defaecation for 5 days. At laparotomy lymphocele, volvulus and ileal stenosis were seen. Lymphocele and stenosis were resected. At second look formation of 2 enterostomies, and at the third look necrotic bowel was resected, leaving 8 cm jejunum, 1cm terminal ileum, and the colon (with ileocecal valve) in situ. The case was discussed in a multidisciplinary meeting and a somber prognosis was concluded. They proposed to withdraw medical treatment but parents disagreed, so treatment continued. One month later, intestinal continuity was restored. At 3 years of age cholecystectomy was performed.

Current status, 7 year old boy, weight 24.5 kg (0.8 SD) and height 120.9 cm (-1.5 SD) with gastrostomy. Partially TPN dependent: enteral feeding 51 ml/kg/day and parenteral feeding 35 ml/kg/day. Medication: zinc sulfate, sodium bicarbonate, Fragmin. Multiple septic episodes, 30 placements of central venous catheters. Occlusion of the left jugular vein. Number of hospital admissions: 57. No elevated liver parameters or other elevated laboratory parameters.

In conclusion, 7 year old boy with total of 9 cm small bowel and colon in situ after intestinal resection for volvulus and lymphocele. Partially TPN dependent, multiple septic episodes and multiple placements of central venous catheters.

QUESTIONS CASE 2

1

Is there medical information missing you need to evaluate this case?

- NO
- YES, namely ____________________________________________________________________________________________

__________________________________________________________________________________________

__________________________________________________________________________________________

2

Would you refer this patient for intestinal transplantation?

- YES (please continue to question 3)
- NO (please continue to question 4)
SUPPLEMENTAL DIGITAL CONTENT 1

3

What are the reasons you would refer this patient?

Please put an X in the box the referral criteria applicable to this patient

| Children with massive intestinal resection          |
| Children with severely diseased bowel and unacceptable morbidity |
| Microvillous inclusion disease or intestinal epithelial dysplasia |
| Persistent hyperbilirubinemia (>6 mg/dl or > 100 µmol/l)     |
| Thrombosis of 2 of 4 upper body central veins           |
| Continuing prognostic or diagnostic uncertainty       |
| Request of the patient and family                   |

Other reasons: ______________________________________________________

____________________________________________________________________

____________________________________________________________________

Please continue to question 5

4

What are the reasons you would not refer this patient?

- Not enough criteria to refer for transplant
- Still enough options for treatment
- Other, ___________________________________________________________

____________________________________________________________________

____________________________________________________________________

5

What would be your treatment proposal?

- Medical treatment: try to increase enteral feeding
- Intestinal lengthening procedure
- Placement on waiting list for intestinal transplantation
- Other, ___________________________________________________________

____________________________________________________________________
Would you list this patient for intestinal transplantation?

- NO
- YES, based on the following criteria (please put an X in the boxes):

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</tbody>
</table>
CASE 3

Boy, born at 37 3/7 week gestational age, birth weight 2350 gr with prenatally diagnosed gastroschisis. Primary closure of the abdominal wall was performed shortly after birth, but on day 1 after primary closure, there were signs of abdominal compartment syndrome. Relaparotomy showed signs of intestinal ischaemia, and the abdomen was closed with a large patch. On day 2 all intestines showed necrosis and all surgical and medical options were discussed in a multidisciplinary session and with the parents. Conclusion was that intestinal transplant was an option. Resection of all bowel from Papilla of Vater up to left part of the colon followed, with 10 cm colon remaining. A gastrostomy was placed concurrently. The boy has been completely dependent on TPN. Six weeks after birth, total bilirubin concentration increased to 187 µmol/l (10.9 mg/dl), and conjugated bilirubin was 140 µmol/l (18.2 mg/dl), caused by incomplete drainage (leading to sepsis and cholangitis). MRCP showed scarring of Papilla of Vater, consequently multiple drains were placed which led to adequate drainage of bile. Neurological development was normal. Parents are very involved in the caretaking and are willing to enter the screening process of possible (combined liver-) intestine transplantation.

In conclusion, 4 month old boy, primary closure of gastroschisis, complicated by necrotic bowel, leaving 10 cm of colon in situ and no small bowel. Completely dependent on TPN, weight 5400 gr (-2 SD). Course complicated by cholangitis for which drainage and 3 septic episodes.

QUESTIONS CASE 3

1. Is there medical information missing you need to evaluate this case?
   - NO
   - YES, namely _________________________________________________________________
     _________________________________________________________________
     _______________________________________________________________
2

Would you refer this patient for:

- Isolated intestinal transplantation (please continue to question 3)
- Isolated liver transplantation (please continue to question 3)
- Combined liver/intestine transplantation (please continue to question 3)
- I would not refer this patient (please continue to question 4)

Please explain your answer: __________________________________________________________

______________________________________________________________________________

______________________________________________________________________________

3

What are the reasons you would refer this patient? Please put an X in the boxes

<table>
<thead>
<tr>
<th>Reason</th>
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Other reasons ______________________________________________________________

______________________________________________________________________________

Please continue to question 5

4

If you would not refer this patient, what are the reasons you would not refer this patient?

- Not enough criteria to refer for transplant
- Still enough options for treatment
- Other, _________________________________________________________________

Continue to question 5
5
What would be your treatment proposal?

- Medical treatment
- Intestinal lengthening procedure
- Placement on waiting list for transplantation
- Other, ________________________________

6
If you would list this patient, what are the listing criteria applicable to this patient?

Please put an X in the boxes

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</table>

Case continued

Current status: 5 ½ years old, history of primary closure of gastroschisis, complicated by necrotic bowel, leaving 10 cm of colon in situ. Weight 14.95 kg (0.1 SD) and height 96.3 cm (-3.0 SD), medication: ranitidine, somatostatine, addamel (trace elements). Still completely dependent on TPN. Multiple septic episodes and central venous catheter placements (7). However on ultrasound, no problems with the major veins.

Chronic pancreatitis with cholecystectomy at 2 years of age. Liver pathology, started 5 months after birth. At the moment septal liver fibrosis, possibly cirrhosis with cholestasis. Laboratory parameters: total bilirubin 66 µmol/l (3.8 mg/dl), conj bilirubin 46 µmol/l (2.7 mg/dl), alkaline phosphatase 457 U/l, γGT 212 U/l, AST 155 U/l, ALT 142 U/l, triglycerides 1.12 mmol/l and PT 12.5 sec.

7
Does this additional information change your decision regarding referral of this patient?

- NO, I would still not refer this patient for transplantation → Continue to question 8
- YES, I would still refer this patient for transplantation → Continue to question 9
I would still not refer this patient for the following reasons:

- Not enough criteria to refer for transplant
- Still enough options for treatment
- Other, __________________________________________________________________________

__________________________________________________________________________________

Please continue to page 35

What are the reasons you would refer this patient? Please put an X in the boxes

<table>
<thead>
<tr>
<th>Reasons</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Children with massive intestine resection</td>
<td></td>
</tr>
<tr>
<td>Children with severely diseased bowel and unacceptable morbidity</td>
<td></td>
</tr>
<tr>
<td>Microvillous inclusion disease or intestinal epithelial dysplasia</td>
<td></td>
</tr>
<tr>
<td>Persistent hyperbilirubinemia (&gt;6 mg/dl or &gt; 100 µmol/l)</td>
<td></td>
</tr>
<tr>
<td>Thrombosis of 2 of 4 upper body central veins</td>
<td></td>
</tr>
<tr>
<td>Continuing prognostic or diagnostic uncertainty</td>
<td></td>
</tr>
<tr>
<td>Request of the patient and family</td>
<td></td>
</tr>
</tbody>
</table>

Other reasons __________________________________________________________________________

__________________________________________________________________________________
I would refer this patient for:

- Isolated intestinal transplantation
- Isolated liver transplantation
- Combined liver/intestine transplantation
- Combined liver/intestine/pancreas transplantation
- Other

If you would list this patient, what are the listing criteria applicable to this patient?

Please put an X in the boxes:

<table>
<thead>
<tr>
<th>Condition</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Small bowel length of &lt;25 cm without an ileocecal valve</td>
<td></td>
</tr>
<tr>
<td>Intestinal failure with high morbidity and poor quality of life</td>
<td></td>
</tr>
<tr>
<td>Congenital intractable mucosal disorder</td>
<td></td>
</tr>
<tr>
<td>Persistent hyperbilirubinemia and signs portal HT or liver dysfunction</td>
<td></td>
</tr>
<tr>
<td>Loss of &gt;50% of standard central venous access sites</td>
<td></td>
</tr>
<tr>
<td>Recurrent life-threatening episodes of sepsis</td>
<td></td>
</tr>
</tbody>
</table>
SUPPLEMENTAL DIGITAL CONTENT 1

You have reached the end of the questionnaire!

We greatly appreciate it that you have taken the time to fill in this questionnaire.

If you have any comments or items that you have missed in this questionnaire:

__________________________________________________________________________

__________________________________________________________________________

__________________________________________________________________________

__________________________________________________________________________

__________________________________________________________________________

__________________________________________________________________________

We would like to thank you again very much for filling in all the questions, we really appreciate your cooperation.

THANK YOU!!!