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# Ontario's Paediatric Referral and Listing Criteria for Small Bowel and Liver- Small Bowel Transplantation

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Version 3.0

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## PATIENT REFERRAL CRITERIA:

The patient referral criteria are criteria for which a Health Care Provider would utilize to refer a patient to a transplant centre for assessment. The criteria identified below are the agreed upon conditions for which a paediatric patient should be referred for intestine transplant assessment. Patients with intestinal failure and one of the following should be referred for assessment for intestine or liver-intestine transplantation

- 1) **Hyperbilirubinemia:** Referral for intestine transplant assessment should be considered for patients with persistent hyperbilirubinemia ( $>80$  mmol/l) for a period of more than 2 weeks that is not associated with sepsis.
- 2) **Thrombosis:** Patients with thrombosis of 2 of 4 upper body central veins or extensive mesenteric venous thrombosis should be referred for transplant assessment.
- 3) **Resection:** Referral for small bowel transplant assessment should be considered for children with massive small bowel resection ( $>75\%$  of small bowel).
- 4) **Disease:** Referral for intestine transplant assessment should be considered for patients with microvillous inclusion disease, intestinal epithelial dysplasia and other congenital diarrheas with high mortality risk.
- 5) **Unmanageable Co-morbidities:** Children with severely diseased bowel and unmanageable co-morbidities including recurrent septic episodes should be referred for transplant assessment.
- 6) **Diagnostic Uncertainty:** Patients with continuing prognostic or diagnostic uncertainty should be referred for transplant assessment.
- 7) **Consent:** Patients or family members may request a referral for transplant assessment.

## PATIENT LISTING INDICATIONS:

Each patient is assessed individually for their suitability for transplantation by the transplant program. The criteria identified below are the agreed upon conditions for which a paediatric patient may be eligible to be waitlisted for intestine transplantation in Ontario. Paediatric listing criteria pertains to those patients  $< 18$  years (up to but not including their 18th birthday).

- 1) **Intestinal Failure Associated Liver Disease:** Listing for transplant may be considered for patients with progressive intestinal failure associated liver disease with persistent elevation of conjugated bilirubin  $>75$ mmol/l or/and signs of portal hypertension, or synthetic liver dysfunction with coagulopathy. The liver disease should be refractory to lipid management strategy (fish oil based emulsion or lipid minimization) that was given for at least 6 weeks.

- 2) **Vascular Access:** Listing for transplant may be considered for patients with loss of  $\geq 3$  standard central venous access sites.
- 3) **Surgical Disasters:** Patients with intractable, severe, abdominal fistulae with poor quality of life are eligible to be waitlisted for transplantation. Listing for transplant may also be considered for patients with extensive mesenteric venous thrombosis.
- 4) **Extensive Local Tumours:** Patients with otherwise unresectable tumors with no or low grade malignant potential (desmoids tumors, lymphangiomas) are eligible to be waitlisted for transplantation.
- 5) **Quality of life:** Listing for transplant may also be considered for patients with intestinal failure with high morbidity and extremely poor quality of life. Listing based on this indication is appropriate for a carefully selected group of patients.
- 6) **ICU Admissions:**  $\geq 2$  admissions beyond the NICU period (and not part of the perioperative period), and that require ventilator support or infusion of inotropes should be considered for listing.

#### **PATIENT LISTING CONTRAINDICATIONS:**

The contraindications for transplantation identified below are the agreed upon criteria for which a paediatric patient would not be eligible to be waitlisted for intestine transplantation in Ontario.

- 1) **Co-Morbidities:** Patients with any co-morbidity that decreases the likelihood of surviving 5 years post-transplant to below 50% or whom which the peri-operative risk is deemed unacceptably high, are not candidates for intestine transplantation.
- 2) **Consent:** Patients who do not want a transplant should not be listed for intestine transplantation.
- 3) **Post-Transplant Care:** Patients with an inadequate or unsafe post-transplant care plan are not candidates for intestine transplantation.
- 4) **Psychosocial Considerations:** Patient non-adherence to therapy is a contraindication to intestine transplantation. Intestine transplantation should be delayed until patients have demonstrated adherence to therapy (compliance with medications) for at least 6 months.
- 5) **Malignancy:** Patients with active malignancy are not candidates for transplantation. In general, patients with a previous history of malignancy must be tumour free before proceeding with transplantation. Most transplant candidates should wait a period of time between successful treatment and transplantation.
- 6) **Pulmonary Disease:** Patients with the following respiratory conditions and severity are not candidates for intestine transplantation:
  - Requirement for home oxygen therapy (Grade C);
  - Uncontrolled asthma (Grade C);
  - Severe cor pulmonale;
  - Severe chronic obstructive pulmonary disease (COPD), pulmonary fibrosis or restrictive disease with any of the following parameters:
    - Best forced expiratory volume in 1 s (FEV<sub>1</sub>) < 25% predicted value;

- $PO_2$  room air  $< 60$  mmHg with exercise desaturation,  $SaO_2 < 90\%$ ;
- $> 4$  lower respiratory infections in the last 12 months;
- Moderate disease with evidence of progression.

**7) Cardiac Disease:** Patients with inoperable valve disease should not be considered for transplant. Patients with severe irreversible (non-uremic) cardiac dysfunction should not be listed for intestinal transplantation alone. Selected patients may be candidates for combined heart–intestinal transplants.

All patients should be assessed for the presence of ischemic heart disease (IHD) before intestine transplantation. The minimum required investigations include history, physical examination, electrocardiogram (ECG) and a chest radiograph. Intestine transplantation is contraindicated in patients with IHD in the following situations:

- Patients with progressive symptoms of angina;
- Patients with a myocardial infarction within 6 months;
- Patients without an appropriate cardiac workup;
- Patients with severe diffuse disease, especially with positive non-invasive tests in whom intervention is not possible and in whom expected survival is sufficiently compromised so that transplantation is not reasonable.

**8) Peripheral Vascular Disease:** Patients with large uncorrectable abdominal aneurysms, severe occlusive common iliac disease, active gangrene or recent atheroembolic events are not candidates for intestine transplantation.

## Version Control:

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