**Appendix: Criteria used to define severe Gaucher’s disease in Ontario**

**Hematological complications (1 or more of the following)**
- Hemoglobin < 85% of lower limit of age- and sex-appropriate normal levels after other causes of anemia, such as iron deficiency, have been treated or ruled out
- Platelet count < $30 \times 10^9$/L on 2 separate occasions at least 1 month apart
- At least 2 episodes of severely symptomatic splenic infarcts confirmed by appropriate imaging of the abdomen

**Skeletal complications (1 or more of the following)**
- 2 acute bone crises severe enough to require hospitalization within 2 years
- Radiographic or MRI evidence of incipient destruction of any major joint, such as hips or shoulders
- Spontaneous fractures confirmed by appropriate imaging studies
- Chronic bone pain, not controllable by administration of non-narcotic analgesics or anti-inflammatory drugs, causing significant loss of time from work or school

**Gastrointestinal complications (1 or more of the following)**
- Evidence of significant liver dysfunction, such as portal hypertension, attributable to Gaucher’s disease
- Significant discomfort owing to enlargement of the spleen or liver

**Pulmonary complications**
- Evidence of clinically significant pulmonary disease owing to Gaucher’s disease

**Systemic complications**
- Growth failure in children: significant decrease in percentile linear growth over 3–6 months