
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

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