Triggers for initiating ACP discussion

At diagnosis of life-limiting condition

See following pages for examples

Life-limiting condition **AND**:

- Family or staff recognise deteriorating patient condition
- Conflict between parents and clinical team regarding use of life-sustaining medical therapy
- 3x unplanned hospital admissions in the past 12 months
- Prolonged hospital admission >3 weeks
- PICU stay ≥1 week
- Multiorgan failure
- Invasive infectious disease
- Initiation of palliative therapy
- Palliative therapy AND new progressing symptoms
- Participation in Phase 1 trial
- Increasing difficulty controlling symptoms

Prolonged or failed attempts to wean off ventilator

“Would you be surprised if this child died within a year?” - answer is “no”

At consideration for transplant (solid organ or bone marrow)

Child or family wishes to discuss ACP
Life-limiting/life-threatening conditions warranting ACP discussion

Bolded conditions require ACP discussion around the time of diagnosis
Unbolded conditions are strongly suggested for early ACP discussion

Malignant
Malignant disease with inevitable fatal outcome
eg DIPG
Actively progressing metastatic disease
Malignant disease with progression on best therapy
Relapsed malignant disease
Malignant disease with predicted outcome ≤40% survival with best treatment
   - Metastatic medulloblastoma
   - Metastatic high risk sarcomas
BMT with stage 4 GVHD

Respiratory
Compromised respiratory status and:
   - Patients with CF considering lung transplant/at the time of transplant
   - Patients with CF with FEV1 < 30%
   - Patients with CF with vent dependence or those ineligible for lung transplant
   - Bronchiolitis obliterans
Central hypoventilation syndromes
Patients who are chronically ventilator dependent

Developmental / genetic
Trisomy 18, 13
Potter Syndrome
Epidermolysis Bullosa
Osteogenesis imperfecta Type 3/4
Severe GMFCS V CP
Other rare chromosomal anomalies with likely poor prognosis
Rett’s Syndrome

Neurological / neurodegenerative / neuromuscular
Progressive neurodegenerative conditions
Muscular Dystrophy
Spinal Muscular Atrophy Type 1
Severe Traumatic Brain injury
Persistent Vegetative State
Batten Disease
Metachromatic Leukodystrophy/ALD
Brain reduction syndromes:
   - Anencephaly
   - Hydranencephaly
   - Lissencephaly
   - Severe schizencephaly
Static encephalopathies
Severe anoxic brain injury
Life limiting/life threatening conditions warranting ACP discussion

Bolded conditions require ACP discussion around the time of diagnosis
Unbolded conditions are strongly suggested for early ACP discussion

**Metabolic**
- Krabbe’s disease
- Hunter’s / Hurler’s disease
- Niemann-Pick disease
- Menke’s disease
- Pompe Disease
- Sanfilippo syndrome
- Tay Sachs disease
- Fabry’s disease
- Sandoff’s disease
  Severe mitochondrial disorder
  Severe metabolic disorders for which bone marrow transplant is a therapeutic consideration

**Renal**
- Neonatal polycystic kidney disease
  Renal failure, not transplant candidate

**Gastrointestinal**
- Short gut syndrome without prospect of curative therapy
- Biliary atresia without prospect of curative therapy
- Multi-visceral organ failure
- Feeding tube under consideration for any progressive or severely disabling neurological condition with no expectation of improvement

**Neonatal**
- Extreme prematurity with concomitant severe BPD, Grade IV IVH, PVL, etc.
- Severe birth asphyxia
- Hypoxic ischemic encephalopathy (moderate to severe)

**Antenatal**
- Any antenatally diagnosed condition likely to be incompatible with life
  Any antenatally diagnosed condition likely to result in shortened lifespan

**Cardiac**
- Discussion of cardiac transplant
- Single ventricle cardiac physiology
- Severe pulmonary hypertension
- Cardiomyopathy: hypertrophic or severe dilated
- Pulmonary atresia (especially if associated with hypoplastic pulmonary arteries)
  Combination of cardiac diagnosis with underlying neurologic/chromosomal diagnosis