

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 \geq 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 $\leq 40\%$ 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

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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