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## **Top Ten Tips Palliative Care Clinicians Should Know About Prognostication in Children**

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## Top Ten Tips Palliative Care Clinicians Should Know About Prognostication in Children

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**Abstract** (100-150 words) 101

Pediatric palliative care (PPC) is different from palliative care for adults. However, conceptualizing PPC remains cumbersome due to the high heterogeneity of often rare diseases, the high diversity of disease trajectories, and the particular difficulty to predict the future of an individual, severely ill child. This article aims to provide an overview and critical reflection of different aspects of prognostication in children with palliative care needs. This includes different diseases from neurology to oncology, from the unborn baby to the young adult, new approaches in treatment, advance care planning, and, most important, communication with the affected child as well as parents.

For Review Only/Not for Distribution

### Introduction:

Prognostication is probably the most demanding task physicians have to learn. Patients and families desire comprehensive information around a life-limiting or life-threatening condition (LL/LTC), which should include a consideration of prognosis. Prognostication in this regard should not be reduced to simply considering the question of “how long” but should include many other factors important to children with serious illnesses and their families. Paul Glare and Christian Sinclair provided important background in their hallmark article on prognostication in medicine;<sup>1</sup> important both for physicians working with adults and for pediatricians, particularly those working with children suffering from LL/LTCs and their families.

Prognostication should be recognized as an important component of care planning, decision-making, and defining goals of care. Uncertainty is an integral part of prognostication, particularly in pediatrics, and should be acknowledged as such. This uncertainty, though, complicates the timing around when to involve palliative care (PC) services, especially in institutions where PC predominantly cares only for those at end-of-life (EOL). Though PC providers understand that PC should be provided independent of prognosis, referring clinicians and parents may conflate PC and EOL care, delaying potentially beneficial support. Whether as an entre to referrals or as a tool to create care plans that align with patient/family values, as accurate an understanding of prognosis as possible is crucial to good care for pediatric patients with LL/LTCs and their families. These top 10 tips for PC physicians aim to illuminate the broad spectrum of prognostication and its impact on the affected child or adolescent, and their family.

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3 **Tip 1: Prognostication is difficult for patients of any age but is particularly difficult in**  
4 **children; its domains include likelihood of cure, anticipated quality of life, feasibility of**  
5 **survival, and length of life.**  
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9 Clinicians may best support communication about prognostication by understanding and  
10 appreciating the multi-layered dimensions of prognostication.<sup>2</sup> The word “prognosis” carries  
11 vastly different meaning for different people. This diversity of definitions warrants clarity of  
12 meaning prior to delving into prognostic disclosures.  
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15  
16 Clinicians are wise to seek parent and patient preferences for the forecasting domains of  
17 prognosis (cure, quality of life (QOL), survival feasibility, length of life). If a parent thinks  
18 prognosis describes *curability* and the provider thinks prognosis means *timeline*, then an  
19 announcement about “weeks to months prior to death” would represent alarming information for  
20 an unbraced parent simply seeking to understand whether their child’s cancer remains curable  
21 with chemotherapy. Conversations about prognosis warrant pacing according to family and  
22 patient readiness,<sup>1</sup> recognizing opportunities to explore additional prognostic domains and build  
23 upon information-sharing during subsequent encounters and growth of the therapeutic  
24 relationship.  
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37 More complex than a binary “cure” or “unable to cure”, prognostication represents the  
38 opportunity to discuss both the current time and the future. Attempts at cure may require travel  
39 or family relocation, extreme symptom burden to the child, or cumulative impact to the child’s  
40 childhood experience of play and relationships. Conversations about prognosis should include  
41 more than physical or body domains and more comprehensively cover topics such ability to play  
42 (known as the work of childhood), to interact with peers and siblings, to engage in  
43 developmentally-relevant tasks, and foster spiritual/existential meaning. While parents often  
44 desire quantification of prognosis in terms of timelines, expanding the conversation to include  
45 additional dimensions of prognosis such as QOL allows clinicians to commit to continued  
46 support beyond a binary “curable/incurable” prognostic content.  
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5 **Tip 2: Prognostication for neonates with LL/LTCs is exceedingly difficult and can**  
6 **inadvertently focus more on function than on infant and family quality of life, reinforcing**  
7 **the value of parallel planning's 'hope for the best and prepare for the worst' approach.**  
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10  
11 Mortality rates are highest for babies around the time of birth and in the first year of life  
12 with 75% of child deaths occurring in this time period.<sup>3</sup> There are also growing numbers of  
13 babies and infants with LL/LTCs with uncertain short and long term prognoses in both high<sup>4</sup> and  
14 low to middle income countries.<sup>5</sup> Diagnoses in this realm include congenital anomalies,  
15 sequelae of prematurity, and birth trauma.<sup>4,5</sup>  
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18  
19 Although there are some diagnoses in babies which will have an inevitable fatal  
20 outcome,<sup>6</sup> there are many more where the disease trajectory and prognoses are uncertain.  
21 Even for those with an expected fatal outcome, defining prognosis in terms of weeks or months,  
22 can be challenging given variability within individual diagnoses. This uncertainty highlights the  
23 need for a more integrative approach to health care including parallel planning<sup>7</sup> where PC and  
24 advance care planning (ACP) are introduced alongside disease-directed care when there is  
25 concern for a LL/ LTC.  
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29 There is a growing understanding of the need for specialty perinatal or neonatal PC  
30 services to work with the existing obstetric and neonatal services in caring for high-risk infants.  
31 Involvement of these PC subspecialists shows some evidence of practice change including  
32 more family centred care<sup>8</sup> but we lack any consensus on the best model of providing this care.<sup>9</sup>  
33 There is also a recognition that this care may require professional education and training  
34 specific to this age group.<sup>10</sup>  
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38 **Tip 3: When discussing prognosis, clinicians should begin by exploring a family's**  
39 **cultural needs and norms, and end with honest, complete, and equitable information**  
40 **delivery.**  
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3 While racial, cultural, and social disparities in prognostic understanding are well-  
4 described among adults with serious illness,<sup>11-14</sup> less is known about pediatric experiences.<sup>15</sup>  
5 This data gap is important because non-white children tend to have potentially inferior EOL  
6 experiences, including increased pain, symptom distress, and intensity of care.<sup>16-19</sup> A 2007  
7 review of PPC studies attributed these disparities to contextual factors (e.g., access to care and  
8 poverty), patient-specific factors (e.g., medical diagnosis), and patient-clinician factors (e.g.,  
9 clinician bias, trust, and quality of information exchange).<sup>15</sup>  
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18 Fortunately, patient-clinician factors are immediately actionable. A study of pediatric  
19 oncologists and parents suggested that ~90% of parents wanted comprehensive prognostic  
20 information, regardless of their race/ethnicity.<sup>20</sup> However, physicians assumed that <33% of  
21 Black and Hispanic parents would want such detail. In a study of parents of children with  
22 advanced neuroblastoma (a disease that is rarely curable), non-white parents were 81% less  
23 likely to understand their child's poor prognosis than white parents.<sup>21</sup>  
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30 These two findings are related; if pediatric clinicians assume diverse families want less  
31 information, they likely will communicate less, in turn limiting the family's opportunity to  
32 understand. Indeed, established barriers to communication among children with serious illness  
33 include foreign language and foreign culture.<sup>19,22,23</sup> Emerging guidelines suggest clinicians must  
34 approach all prognostic communication first with curiosity: assume nothing and deliberately  
35 explore a family's cultural needs, norms, and style.<sup>23</sup> When families are non-English speaking,  
36 communication must be done with a certified medical interpreter. Next, clinicians should deliver  
37 comprehensive prognostic information compassionately and honestly. Variations in approach  
38 should be tailored to the family's endorsed preferences to improve prognostic understanding for  
39 all patients and families.  
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54 **Tip 4: Though adolescent and young adult patients with cancer desire high-quality**  
55 **prognostic communication from the time of diagnosis, mutual protectionism between**  
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3 **child and parent may delay external prognostic discussions independent of internal**  
4 **prognostic awareness.**  
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6  
7 Adolescent and young adult (AYA) patients with cancer consider prognostic information  
8 important and more extensive prognostic disclosure is associated with improved patient well-  
9 being and higher trust in the oncologist.<sup>24</sup> Three-quarters of AYAs expressed that having a  
10 precise, numeric understanding of prognosis was important at the time of diagnosis and this  
11 percentage increased over the first year.<sup>25</sup> Honest communication about prognosis in advanced  
12 cancer, including near the EOL, allows for open exploration of patient and family hopes, worries,  
13 and decisions about medical care.<sup>26</sup>  
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22 Unfortunately, open prognostic discussions and associated larger goals of care may be  
23 hindered by mutual pretense, wherein both parties know information but do not openly  
24 acknowledge it with each other.<sup>27</sup> Parents may seek to avoid open discussions about prognosis  
25 with their child to protect or shield them from harm and conversely some children may avoid this  
26 open communication to similarly protect their parents.<sup>28,29</sup> However, inadequate prognostic  
27 communication in AYAs with cancer may result in anxiety and fear at the EOL<sup>30</sup> and lingering  
28 regret in parents following the child's death.<sup>31</sup> In a large study of bereaved parents, over one-  
29 quarter of those that did not discuss prognosis with their child regretted this decision; parents  
30 were more than three times as likely to express regret if the child was a teenager.<sup>32</sup> Clinicians  
31 must consider personal, familial, and cultural differences in prognostic disclosure, while  
32 engaging in longitudinal, compassionate exploration of both parent and AYA preferences.<sup>26</sup>  
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47 **Tip 5: Prognostication in advanced pediatric cancer remains difficult and, unlike in adult**  
48 **medical oncology, prognostic and functional scales offer little to no useful guidance.**  
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50 While several prognostic scales have been developed and tested in adult populations  
51 (e.g., Glasgow Prognostic Score, Palliative Performance Scale, Palliative Prognostic Score,  
52 Palliative Prognostic Index, Prognosis in Palliative Care Study (PiPS)), similar scales in the  
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3 pediatric cancer population are lacking.<sup>33</sup> Some pediatric scales were developed or tested  
4 across a variety of LLCs so performance in oncology may be unknown. As well, some scales  
5 focus on when to refer a child and family to PC services rather than on prognostication, though  
6 the two are related.  
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11 In adults, a common prognostic approach is the surprise question: would you be  
12 surprised if this person died in the next 12 months.<sup>33</sup> In pediatrics, the surprise question may be  
13 more accurate than has been demonstrated in adult populations.<sup>34</sup> For children with cancer,  
14 sensitivity of the surprise question was 100% but specificity was 33%, while specificity was 81%  
15 and 70% for children with neurological or congenital illnesses respectively.<sup>34</sup> The Paediatric  
16 Palliative Screening Scale (PaPaS) was developed and validated to determine the need for PC  
17 in children with any LLC, but incorporates the surprise question with a six-month window.<sup>35</sup> The  
18 PaPaS has demonstrated feasibility and utility in a mixed population to identify PC needs.<sup>36</sup>  
19 Finally, specific to children who received an allogeneic hematopoietic cell transplant (HCT) and  
20 required intensive care unit (ICU) admission, risk of mortality during the admission increased  
21 when there was pre-existing renal comorbidity, cytomegalovirus seropositivity, less than 100  
22 days from HCT to ICU admission, acute myelogenous leukemia as the underlying condition, and  
23 a high Pediatric Risk of Mortality 3 (PRISM-3) score.<sup>37</sup> Outside of the specific situation of HCT,  
24 the surprise question and PaPaS may offer some guidance for prognostication in pediatric  
25 cancer, but more research is needed.  
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45 **Tip 6: Several chronic conditions, including Trisomy 13/18, can have wide variations in**  
46 **prognosis depending on the aggressiveness of treatments utilized.**  
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49 Trisomy 13 (T13) and 18 (T18) are genetic diagnoses that confer multiorgan system  
50 anomalies including cardiac malformations and neurologic impairment. T13, also called Patau  
51 syndrome, occurs in approximately 1.68 per 10,000 live births and T18 (Edwards syndrome)  
52 occurs in approximately 4.08 per 10,000 live births.<sup>38</sup> Despite historic teaching that these  
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3 conditions are “uniformly fatal” or “incompatible with life”, multiple large-scale studies have  
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5 recently demonstrated that a proportion of children with T13 and T18 will live for years. One  
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7 large international cohort study found that for infants with T13 born live, median first-week  
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9 mortality was 48%, median first-year mortality was 87%, and cumulative 5-year survival was  
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11 7%.<sup>38</sup> That same study found a median first-week mortality of 42% for T18, median first-year  
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13 mortality of 88%, and cumulative 5-year survival of 7.7%. In a study of patients born in Ontario,  
14  
15 Canada over 21 years, Nelson et al. found a one-year survival rate of 19.8% in T13 and 12.6%  
16  
17 in T18 and that among those infants with T13 and T18 alive at 6 months, 50.5% and 60%  
18  
19 respectively survived to 10 years.<sup>39</sup> Importantly, cardiac and neurologic diagnoses or congenital  
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21 anomalies in more than one organ system did not confer worse survival in T13 or T18.<sup>39</sup>  
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24 Evidence is mounting that surgical and medical interventions may further improve  
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26 survival rates in children with these conditions.<sup>40</sup> Yet despite an improved understanding of  
27  
28 long-term survival in T13 and T18, parents continue to report poor quality medical  
29  
30 communication regarding prognosis and decision making for these children.<sup>41</sup> An  
31  
32 interdisciplinary approach is warranted in the care of children with T13 and T18, integrating PPC  
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34 clinicians prenatally and continuing longitudinally, in order to ensure parental counseling that is  
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36 accurate and attentive to each family’s unique goals and values for the child’s entire length of  
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38 life.<sup>42</sup>  
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43 **Tip 7: In children with spinal muscular atrophy (SMA) and other genetically determined**  
44 **life-limiting conditions, gene therapy is a game changer in pediatric palliative care since**  
45 **it prolongs life substantially and leads to new questions regarding ethics, health care**  
46 **costs, and uncertainty of life expectancy.**  
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51 Not all rare conditions are life-limiting, but a large majority of children receiving PPC  
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53 suffer rare diseases. The Food and Drug Administration (USA) and the European Medicines  
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55 Agency (EUROPE) implemented strategies like the Orphan Products Development to enable  
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3 development of drugs for rare diseases. These drugs, biological products and techniques will  
4 change care for children with LLCs and raise ethical questions. However, it remains unclear  
5 when to stop the treatment if gains have not been achieved. Nusinersen and the other gene  
6 therapies will change the way of care.<sup>43</sup> They provide palliation but no cure. In parallel, re-  
7 imbursement may challenge healthcare systems as well as families.  
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14 SMA, an autosomal recessive disorder, is the most common genetic cause of infant  
15 mortality in the USA (Carrier rate 1:50; incidence 1 in 10,000 births). SMA type I counts for 60%  
16 of all cases. Typically, newborns show muscle weakness (never able to sit independently) and  
17 respiratory impairment. Without extensive treatment 70% die before their second birthday.  
18 Children with SMA type II can sit without support, but cannot stand or walk independently. The  
19 majority of patients live into early adulthood.  
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26 Patients with SMA have a mutation in the survival motor neuron (SMN) 1 gene that leads  
27 to motor neuron loss; nusinersen (Spinraza, Biogen), a breakthrough gene-therapy treatment  
28 approved in late-2016, increases SMN protein production leading to near normal motor  
29 development. In the USA, direct medical treatment costs are about \$ 324,410/year<sup>44</sup> and  
30 reimbursement in Europe depends on the country. Meanwhile, universal newborn screening  
31 tests are available in several countries that allow treatment with gene therapies before  
32 symptoms occur.  
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41 While celebrating the medical breakthrough, modern medicine transforms many serious  
42 diseases into chronic conditions requiring people to remain a “patient” forever (e.g. HIV disease;  
43 organ-allo-transplantation). That leads to a lifelong balancing act between desired and  
44 undesired treatment effects, burdens, and gain of therapy. New challenges are lifelong need for  
45 (financial) support, the desire to live independently, to participate in the workforce, and to have  
46 an own family. In SMA I/II patients receiving gene therapy, several new uncertainties remain  
47 unanswered: Who benefits and for how long? What are the long-term sequelae of treatment?  
48 Will the treatment improve language acquisition and cognitive outcomes?<sup>45</sup>  
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5 **Tip 8: Markers of declining prognosis include failure of the gastrointestinal system**  
6 **(feeding intolerance, retching, and pain), increasing hospital admissions without the**  
7 **return to baseline level of functioning, and a diminishing quality of life as defined by the**  
8 **family.**  
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14 Severe neurologic impairment (SNI) describes a group of disorders of the central  
15 nervous system arising in childhood, resulting in motor and cognitive impairment and medical  
16 complexity, typically requiring much assistance with activities in daily living.<sup>46</sup> Up to 40% of  
17 children cared for by PPC teams have SNI.<sup>47</sup> The vast majority live with medical technology.  
18 Because SNI comprises myriad disease states, patterns of decline and length of life are highly  
19 variable; prognostic uncertainty and limited empirical evidence complicate how to best counsel  
20 their families.<sup>47,48</sup>  
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28 Although many children with SNI have static encephalopathy, physical decline across  
29 SNI conditions can mirror that observed in adult neurologic disorders such as dementia or  
30 following traumatic brain injury. Feeding intolerance manifested by retching, vomiting or pain  
31 with feeds may occur, prompting reassessment of feeding goals.<sup>49</sup> Worsening seizures, central  
32 or obstructive apnea, autonomic dysfunction and neurogenic bowels and bladder; increased  
33 global pain and discomfort; and changes in consciousness may also occur over time.<sup>49</sup>  
34 Restrictive lung disease due to neuromuscular weakness and scoliosis is generally progressive,  
35 with rate of decline accelerated by recurrent pneumonia and slowed by the use of positive  
36 pressure ventilation and aggressive respiratory care.  
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47 Physical changes may be gradual or occur abruptly and then be followed by a period of  
48 stability for years, complicating prognostication. Additionally, children with SNI are at risk for  
49 cascading symptoms—feeding intolerance may lead to increased visceral hyperalgesia and  
50 autonomic dysfunction, worsening seizures and exacerbating hypertonicity, for example—and  
51 careful attention must be paid when assessing symptoms in order to treat perceived  
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suffering.<sup>47,49</sup> For these reasons, children with SNI and their families benefit from ongoing comprehensive PC across all phases of their lives.<sup>47</sup>

**Tip 9: There are times when the most accurate prognostic approach simply acknowledges uncertainty; rather than offering a specific prognosis, PPC teams should support parents and children as they journey through the proverbial wilderness.**

Prognostication is difficult in PPC. Many of the conditions encountered are rare or even one-of-a-kind. Even in conditions in which there is considerable knowledge about the group as a whole, there can be great phenotypic variability. Technological advances create additional challenges. In this setting, predicting any individual child's lifespan can be fraught and there is evidence to suggest that clinicians are often wrong when making predictions about how long a patient may have to live.<sup>50</sup>

Parents often report that uncertainty is one of the most challenging aspects of caring for a child with a LLC.<sup>51</sup> Clinicians can feel an overwhelming need to help provide some certainty by attempting to predict the future. Even if the certainty is of a bad outcome or a short lifespan, there is a sense that knowing this can help families prepare and make important decisions about how to spend the time they have left together.<sup>51</sup> However, a life of suffering that continues for longer than anticipated or a life cut suddenly short can create a great deal of distress. It may that the real task for those who work in PC is to work with humility and to acknowledge and help families navigate uncertainty. Parallel planning may be a helpful approach,<sup>52</sup> with various potential scenarios are explored and plans put in place for each of these.

**Tip 10: Anticipatory and post-death grief varies widely for children and parents; the dual process model of loss-oriented and restoration-oriented tasks can provide a cognitive framework for clinicians' understanding of the family's perspective or actions.**

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3 Anticipatory grief starts from diagnosis or recognition of a LL/LTC<sup>53</sup> as the family adjusts  
4  
5 to a different future than the one they had previously envisioned for their child. However the  
6  
7 experience and expression of anticipatory and post-death grief varies widely. One proposed  
8  
9 approach to deal with grief is called the 'dual process model'.<sup>54</sup> In this schema, loss-oriented  
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11 stressors and restoration-oriented stressors are held simultaneously by families. These  
12  
13 stressors are confronted or avoided as needed given the situation at hand. For example, in the  
14  
15 context of anticipatory grief, a parent might at times focus on planning for their child's return to  
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17 school, whilst at other times focus on planning their funeral.  
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20 From time to time patients' and/or their families' words and actions suggest that perhaps  
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22 they have not understood or accepted prognostic information given to them. However this is not  
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24 necessarily the case. Misinterpreting these cues can lead to misplaced emphasis on cure  
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26 directed therapy, prolonging the child's life at all costs or inappropriate emphasis on "getting the  
27  
28 message home". Either of these approaches are likely to increase distress for the patient,  
29  
30 parent, and clinician.  
31  
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33 Awareness of the dual process of grief can help the clinician explore and understand  
34  
35 better the thoughts and feelings of the family member. The clinician is then able to better pace  
36  
37 the provision of prognostic information in response to cues from the family member with benefits  
38  
39 including mutual understanding, increased trust and satisfaction with communication,<sup>55</sup> and  
40  
41 reduced symptom burden.<sup>56</sup> Awareness of the dual process model can also be helpful when  
42  
43 facilitating ACP discussions: allowing exploration of hopes as well as fears, supporting the  
44  
45 family to anticipate and plan for both positive and negative events, and naming the inherent  
46  
47 uncertainty to facilitate coping.<sup>57</sup>  
48  
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50

## 51 **Conclusion:**

52 Prognostication has many different dimensions. In PPC, this is of particular relevance.  
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54 Discussing prognosis with patients and families may provide the opportunity to support patients  
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and families as they actively participate in the process of defining goals of care and treatment, allowing autonomy and ideally enhancing resilience for everyone involved.

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