Advance Care Planning: practicalities, legalities, complexities and controversies

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ABSTRACT

Increasing numbers, complexities and technology dependancies of children and young people with life-limiting conditions require paediatricians to be well prepared to meet their changing needs. Paediatric Advance Care Planning provides a framework for paediatricians, families and their multidisciplinary teams to consider, reflect and record the outcome of their conversations about what might happen in the future in order to optimise quality of clinical care and inform decision-making. For some children and young people this will include discussions about the possibility of death in childhood. This may be unexpected and sudden, in the context of an otherwise active management plan or may be expected and necessitate discussions about the process of dying and attention to symptoms. Decision-making about appropriate levels of intervention must take place within a legal and ethical framework, recognising that the UK Equality Act (2010) protects the rights of disabled children and young people and infants and children of all ages to the same high quality healthcare as anyone else.

INTRODUCTION

This article sets out to:

▸ Describe paediatric Advance Care Planning (pACP) and its various elements;
▸ Discuss the evidence to support pACP;
▸ Give pragmatic guidance about pACP for paediatricians based on extensive personal experience;
▸ Discuss the tricky areas of prognostication and decision-making about appropriate levels of intervention that underpin pACP.

WHAT IS PAEDIATRIC ADVANCE CARE PLANNING AND WHY DO IT?

Advance Care Planning (ACP) has different meanings depending on perspective of country and legislation. In broad terms it is a description of the process of discussing issues and planning ahead in anticipation of a change in condition in future. The components of pACP for some families will include discussions about the possibility of premature death. If these conversations are not had, families will miss out on opportunities to state their wishes for aspects of care and interventions, including place of care and preferred place of death. Clinician-led discussions about the possibilities of dying and death in advance may help families to communicate among themselves about what may otherwise have remained unspoken, may help with the grieving process and improve realistic hope and resilience.

WHAT IS THE EVIDENCE TO SUPPORT ADVANCE CARE PLANNING IN PAEDIATRIC CLINICAL PRACTICE?

The prevalence of life-limiting conditions in children is increasing, bringing an increasing clinical need to recognise when dying and death may be possibilities, so that appropriate discussions can be had with families. Balanced against a dearth of randomised, controlled trials in the area of pACP decision-making about appropriate levels of intervention and end-of-life care for children, there are plenty of qualitative and descriptive publications evidencing good clinical practice. The Royal College of Paediatrics and Child Health is currently updating their guidance on decision-making for children with life-limiting conditions and the National Institute for Health and Care Excellence (NICE) in England is planning a clinical guideline on end-of-life care in children, both eagerly anticipated; it will be interesting to see how much evidence their rigorous processes identify.

One key message from the literature is without doubt: planning and care for children at risk of premature death needs to improve. A systematic review (2013) reported on three pACP programmes in the USA, two developed from adult programmes. These involved discussions with families in advance, involvement of chaplains and recording of ‘advance directives’. Preliminary findings were that pACP can be implemented and was perceived as helpful. A randomised controlled trial specific to HIV-infected adolescents showed pACP to trigger positive emotional experiences in young people and their carers with less negative reactions when compared with those in the study’s control arm not receiving pACP also a better communication experience and marginally improved certainty about decisions in the intervention group. A subsequent randomised controlled trial of family centered pACP in young people with cancer concluded that those receiving pACP were significantly better informed about end-of-life decisions and the young people were more likely to allow family members to ‘do what is best at the time’ (100%) compared with the control group who did not receive pACP (62%). Those in the pACP group were significantly more likely to limit treatments, knowing that this accorded with the young person’s own wishes.

Consistently across studies young people and their carers wanted more information and to be involved in decision-making. Reported barriers to pACP included lack of many children’s capacity...
to be involved in decision-making themselves, the high degree of
prognostic uncertainty, unrealistic expectations of parents,
doctors’ worries about removing hope and complexity of the
social environment. In one study 71% of doctors questioned thought pACP happened too late. No studies compared
frequency of home deaths in children with and without pACP,
but two studies suggest home deaths may be more frequent
where pACP had occurred.

A study on Birmingham’s paediatric intensive care unit cap-
tured views of eight consultants and six senior nurses. They
felt that children were ‘enduring’ intensive care which they
believed to be ‘futile’ and expressed the view that pACP was
lacking, but if implemented would improve care for children
with life-limiting conditions.

ELEMENTS OF PAEDIATRIC ADVANCE CARE PLANNING
Emergency healthcare planning
It is good clinical practice to provide for families with children
with complex healthcare needs a written care plan that includes
information about their diagnoses, active health concerns and
any emergency scenarios that can be predicted to arise, includ-
ing written guidance about what to do from presentation in the
community through to the hospital emergency department (see
figure 1). This is promoted in the UK through ‘Together for Short Lives’ (http://www.togetherforshortlives.org.uk) who have
many helpful supporting resources for families and profes-
sionals. A national survey of UK paediatric intensive care units
found wide variation in availability and nature of formal pACP
documents and called for development of national policy
informed by review and evaluation of existing documents and
current practice in pACP.

In 2011, the Department of Health (England) funded a col-
laboration of paediatricians representing general, community,
emergency, disability and palliative care paediatrics who together with the Council for Disabled Children agreed core
principles for Emergency Health Care Plans. It was acknowl-
edged that localities required their own easily recognisable
formats for such Plans across services, including Ambulance
Services. The outputs from this work, including e-Learning
resources, regional exemplar templates, a parent information
leaflet and video exemplars of enacted difficult conversations
with families can be found at: http://www.councilfordisabledchildren.org.uk/ehp.

Advance statement of wishes and preferences
Fraser and team from Bristol have led on this work in the UK. Practical templates, flow charts and resources are accessible at http://www.togetherforshortlives.org.uk/professionals/external
resources/

Statement about discussions with the child and parents
about appropriate levels of intervention and the
circumstances in which these will apply
This is the trickiest area, as there is a need for the paediatrician
to have discussed the issue of prognosis with the child, their
parents or guardians. Prognostication can be extremely difficult; it falls into two broad groups:

- **Risk of sudden and unexpected premature death in childhood**
  - There is evidence that children with long-term conditions
    are at higher risk than others of sudden and unexpected
    death. Paediatricians are not universally confident about
discussing this risk with families but increasingly
    need to see this as part of their role.

- Paediatricians may be familiar with the concept of Sudden
  Unexpected Death in Epilepsy. Recent evidence suggests
  that it is the complexity of the child’s disabilities rather
  than the epilepsy itself that is the main risk factor for
  sudden death. There is also evidence especially from
  support groups (eg, http://www.epilepsyaction.org.uk;
  http://www.sudep.org) that families deal better with their
  loss and grief if there had been a discussion about risk of
  death in advance. Thus paediatricians caring for children
  with long-term conditions, especially those with complex
disabilities, need to consider the possibility of death and
discuss risks openly with families. Discussing the possibility
of death does not mean that active treatment should cease.
It should never be assumed that early death is inevitable
and pACP always needs to embrace a range of possible
outcomes.

- Clinicians should be mindful of the improved survival
  opportunities for even those with complex disabilities, who
  may not follow the journey predicted for their condition in
  the published literature and who may respond well to
  intensive care to tide them over episodes of acute illness.
The multifaceted needs of the child should continue to be
addressed, based on best evidence, aiming always for the
best possible quality of life and fullest possible participation
in everyday activities.

- The mode of death in the sudden, unexpected group is as
  it says on the tin and can occur at any time, in any setting,
much usually at night. Families need to be sensitively made
aware of this, while setting the risk in context and not
making them overanxious.

- **Risk of gradual decline, dying and death**
  - Identifying this group requires the paediatrician to be alert
    for ‘red flags’ that the dying process may be imminent or
    occurring. Some suggested ‘red flags’ from personal experi-
    ence are listed in figure 2.

  - Children at risk include those:
    - for whom active treatment is no longer effective and/or
      the disease process is progressing;
    - with an underlying static medical condition, usually neuro-
      logical, chromosomal or genetic, where secondary complications
develop their own momentum, unchecked by best efforts of
medical interventions.

  - The conversation with families about risks of dying and
death for this group is necessarily different, as the clinician
  is sensitively letting the family know that death is expected,
  although the precise timing of death cannot be predicted
  and even the frailest children continue to surprise us with
  their remarkable resilience.

  - It should be acknowledged by the clinician that resuscita-
tion attempts are likely to be unsuccessful and that further
active treatments are likely to be futile; both may be bur-
densome for the child and distressing for the family. This
needs to be recorded on the family held care plan so that
the lead clinician at all times is apprised of what has been
discussed and with whom.

  - Having a conversation with a family about the possibilities
of dying and death is difficult for all involved and requires
careful thought and preparation. Some tips from personal
experience to consider are given in figure 3.

  - If death is inevitable in the clinical judgment of the lead
clinician and a decision is made that further life-sustaining
  treatment is inappropriate and would be burdensome,
there are a number issues to take into account, including in
any pACP documentation (see figure 4).

Horridge KA. Arch Dis Child 2015;100:380–385. doi:10.1136/archdischild-2014-305945 381

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Arch Dis Child: first published as 10.1136/archdischild-2014-305945 on 1 October 2014. Downloaded from http://adc.bmj.com/ by guest. Protected by copyright.
The mode of death for this group is more likely to be gradual, most often with significant respiratory failure compounded by atelectasis and variable infection all of which may contribute to symptoms that will need to be addressed to ensure comfort at all times and explained to the family. Families may get upset about the length of time that the dying process can take. Being alongside them at this difficult stage of their child’s journey and continuing to offer explanation and support is an important role of the clinical team and much appreciated by families.

Decision-making frameworks
All decision-making must be within an ethical and legal framework. In the UK clinicians are bound to follow the 2010 General Medical Council guidance on treatment and care towards the end of life that includes specific guidance on decision-making for children and young people23 and also the Equality Act (2010)24 that includes disability and age among the protected characteristics that must not be used as a reason to treat some people worse than others. The Children Act (1989 and 2004) applies throughout the UK and includes a seven-point welfare checklist that must underpin any decisions made in the courts or best interests decisions about children and young people up to 18 years old.25 In England and Wales those aged 16 years or more come under the 2005 Mental Capacity Act (MCA) that defines tests of capacity and sets out a minimum nine-point checklist for estimating the best interests of a person who lacks capacity.26

The process of decision-making as well as the actual decision should be written in the medical record and also in any family held care plan, so that in the event of a healthcare emergency it is clear to all who may be involved as to what has been discussed and decided, in the child’s best interests.

Emergency health care planning
(http://www.councilfordisabledchildren.org.uk/ehp)
- Include demographic data about child or young person and their family
- List active health concerns and diagnoses
- List current treatments or signpost to where these may be found
- Predict specific scenarios that may arise for the individual child or young person and practical guidance to follow, from community setting through to hospital emergency department
- Give details of involved clinicians, including who to contact in an emergency

Advance statement of wishes and preferences29
- Include specific wishes and preferences of the child or young person and their parents or carers
- These may include:
  - specific people to contact in specific circumstances
  - preferred place of care
  - any other details that are important to the child or young person and family that they want to be transparently available in a healthcare emergency or in case of change in condition

Statement about discussions with the child and family about appropriate levels of intervention and the circumstances in which these will apply
- For most children and young people this will state that assessment and management should follow Advanced Paediatric Life Support (APLS) guidance at all times, including full resuscitation and intensive care if required. This protects the right of a disabled child to receive the same high standard of healthcare as anyone else and mitigates for a clinician who may be unfamiliar with them from making decisions based on their own preconceived ideas about their quality of life22.
- For a minority of children and young people where death is predicted soon as a consequence of a progressive disease process or treatment failure, or where their condition is so very frail as to make resuscitation attempts unlikely to succeed, a statement to the effect that the child or young person should be allowed a natural death when their time comes should be recorded. Some regions require specific documentation of such decisions in a locally recognised format, for example, a ‘Do Not Attempt Cardiorespiratory Resuscitation’ order or ‘Personal Resuscitation Plan’.

For people aged 18 years and over, elements of UK capacity legislation can support Advance Care Planning for those who have capacity for specific decisions, so that their wishes can be taken into account by the lead clinician should they lose capacity in the future20
- Advance Decision to Refuse Treatment
- Appointment of a Personal Health and Welfare Lasting Power of Attorney
- Support with best-interests decision-making from the Court of Protection

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**Figure 1** Component elements of paediatric Advance Care Planning.
Figure 2  Red flags that dying and death are significant possibilities in children and young people who are medically frail.

**Increased frequency of:**
- time spent unwell
- biomarkers indicating relentless progression of disease process despite best treatment
- severe infections, taking longer to respond to treatment
- admissions to hospital and/or to paediatric intensive care
- feeding difficulties and swallowing dysfunction
- need for suction to clear upper airway due to decreased ability to clear own secretions
- treatment-resistant seizures
- agitation
- pain, which may be non-specific and no cause may be identified despite thorough clinical assessment
- changes in sleep, behaviour and personality
- ‘control centre malfunction’ as evidenced by intermittent or continuous autonomic dysregulation with variations in temperature, breathing, skin colour and sweating that are not explained by medically identifiable causes such as sepsis

Figure 3  Tips to consider when planning a conversation about the possibilities of dying and death of a child or young person.

- Always actively listen to the child or young person, check what they know and understand at every step
- Always actively listen to parents and carers, mindful that each may have a different view and have different needs in terms of how information is shared, understood and assimilated
- Seek permission to share difficult information and check that the setting and circumstances are as supportive as can be
- Give permission for others to be present as needed e.g. a grandparent, advocate or friend
- Don’t be afraid of silence. This is an important component of active communication, giving time for reflection and assimilation of information
- Repeat important information and follow up in writing for the family and key involved professionals with consent
- Consider the needs of brothers and sisters and other family members and offer support in communicating with the wider family
- Involve the multidisciplinary team: general practitioner, therapists, specialist children’s nurses etc. It is essential for the team around the child to be aware, with consent, about the information that has been shared to avoid misunderstandings
- Ensure prompt follow up, which may be with a different member of the team, to check understanding of what has been said and to offer support
- Avoid absolutes. Trying to predict length of life and timing of events is fraught with difficulties although many families will push for this. Remain humble about what is known and what is not
- Assure the family and carers of ongoing attention to symptoms and support
- Be proactive in predicting symptoms and scenarios that might arise and ensure there is a robust care plan in place and that any medication that may be required is easily accessible and available should an emergency arise
- Remember that it is discriminatory to put in place a ‘Do not attempt cardiopulmonary resuscitation’ order when an arrest is not an event that is anticipated.
- Discussing the possibility of sudden or ‘unexpected death’ does not mean that active treatment needs to stop. The child or young person can still receive full resuscitation and intensive care as required for as long as this is appropriate for them and has a chance of being successful. A positive statement about this should be included in the medical record and family-held care plan to protect the right to full and active treatment.
- Supporting families through uncertainty, where there is a range of outcome possibilities, although tricky for clinicians is important for families, who value supportive honesty above all else
- Being there alongside the child or young person and their parents and carers on their journey is as important as medicines in the overall therapeutic milieu and should not be undervalued
- Be aware of the stages of the grieving process that families may encounter when they receive difficult news, including disbelief, anger, guilt, bargaining, searching, fighting, sadness and in time, acceptance
- Be mindful of one’s own feelings and emotions as a professional and do not be afraid to seek supervision and support. We are not always good at looking after ourselves as practitioners but we are no good to those we serve if we neglect to do so
Advance Care Planning with and for children under 16 years of age

In UK law, children under 16 years of age are presumed to be incompetent to make their own decisions, although legal mechanisms exist that allow them to demonstrate their competence for specific decisions, for example, Fraser guidelines and Gillick competence test.27 While legally competent to make some decisions about themselves, young people face the extra challenges of adolescence including emerging and variable maturity and concrete thinking. The paediatrician, mindful of these complexities, should refer to the Children Act (1989 and 2004) welfare checklist25 to guide any decision made by, with or on behalf of children and young people in their best interests.

Advance Care Planning with and for 16-year-olds and 17-year-olds and transition issues

There is a presumption in law in England and Wales that 16–17-year-olds have capacity to consent to treatment and can refuse some treatments. For those assessed to lack capacity, a person with parental responsibility, usually a parent, should be consulted by the clinician who will make a decision in the young person’s best interests. Also, the Children Act (1989 and 2004) allows a parent to consent to treatment as for a younger child. It may be necessary to seek legal advice in case of uncertainty or disagreement.

In Scotland, 16-year-olds are considered to have capacity and can consent or withhold consent for themselves in the same way as a person 18 years of age or over in England and Wales under the MCA. For a young person in Scotland assessed to lack capacity for specific decisions, a power of attorney may be appointed by application to the Office of the Public Guardian (Scotland).28

In North-East England, a collaboration of palliative care physicians and paediatricians produced an all-age suite of support tools for decision-making and recording that is also being adopted in other regions.29 This overcomes difficulties that otherwise might arise at transition between children’s and adult services, where ‘best interests’ may be differently defined.

Whatever process is adopted, it is essential to be mindful of the MCA best interests process for all young people over 16 years of age who lack capacity for a specific decision at a specific time and in specific circumstances.

CONCLUSIONS

In the face of increasing numbers of children with life-limiting conditions, paediatricians need to be well prepared to plan...
ahead for clinical scenarios that might arise, to recognise when unexpected or expected death may be possible and to discuss risks openly with families. This allows the child where possible and their parents and carers, to express and record their wishes and preferences and for conversations to be had between the family and multidisciplinary team about appropriate levels of interventions. Increasing the number of conversations about dying and death with families and improving recording of wishes and decision-making using a pACP framework will facilitate future research to explore pACP more thoroughly, with the hope of making the most difficult journey of all for families a little more bearable.

Competing interests None.

Provenance and peer review Commissioned; externally peer reviewed.

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