

# Compassionate deactivation of ventricular assist devices in pediatric patients



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## KEYWORDS:

pediatrics;  
ventricular assist;  
palliative care;  
mechanical support;  
quality of life

Despite greatly improved survival in pediatric patients with end-stage heart failure through the use of ventricular assist devices (VADs), heart failure ultimately remains a life-threatening disease with a significant symptom burden. With increased demand for donor organs, liberalizing the boundaries of case complexity, and the introduction of destination therapy in children, more children can be expected to die while on mechanical support. Despite this trend, guidelines on the ethical and pragmatic issues of compassionate deactivation of VAD support in children are strikingly absent. As VAD support for pediatric patients increases in frequency, the pediatric heart failure and palliative care communities must work toward establishing guidelines to clarify the complex issues surrounding compassionate deactivation. Patient, family and clinician attitudes must be ascertained and education regarding the psychological, legal and ethical issues should be provided. Furthermore, pediatric-specific planning documents for use before VAD implantation as well as deactivation checklists should be developed to assist with decision-making at critical points during the illness trajectory. Herein we review the relevant literature regarding compassionate deactivation with a specific focus on issues related to children.

J Heart Lung Transplant 2016;35:564–567

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Despite greatly improved survival in pediatric patients with end-stage heart failure through the use of ventricular assist devices (VADs), heart failure ultimately remains a life-threatening disease with a significant symptom burden. With increased demand for donor organs, liberalizing the boundaries of case complexity, and the introduction of destination therapy (DT) for VAD implantation in children, more children can be expected to die while on mechanical support.<sup>1–5</sup> Even for patients who survive, common adverse events, such as stroke, bleeding, infection and end-organ failure, can greatly reduce quality of life for these children, and lead to discussions concerning the desirability of ongoing life-sustaining therapies.<sup>6–10</sup>

In the United States it is both ethical and legal to withdraw life-sustaining therapies, including VAD support, at the request of an adult patient or his/her surrogate who perceives the burdens of therapy outweigh the benefits, a point of view repeatedly reinforced in expert reviews and consensus guidelines from professional societies.<sup>11–17</sup> In contrast, similar guidelines on the ethical and pragmatic issues of compassionate deactivation (CD) of VAD support in children are strikingly absent. Herein we review the relevant literature regarding CD with a specific focus on issues related to compassionate discontinuation of VAD support in children.

## Case report

An 11-year-old boy with a history of dilated cardiomyopathy and orthotopic heart transplantation in infancy presented to the pediatric heart transplant clinic with severe,

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symptomatic (New York Heart Association Class IV) biventricular heart failure without evidence of rejection or graft vasculopathy. Despite maximal medical therapy the patient continued to decompensate, and eventually underwent biventricular VAD implantation with Thoratec pediatric VADs as a bridge to re-transplantation. The course was complicated by inadequate ventricular decompression leading to significant left atrial hypertension, acute kidney injury and progressive respiratory failure. Despite several interventions and adjustments to the devices, stable hemodynamics could not be achieved. On Day 59 post-operatively, the patient had a large embolic stroke in the left middle and anterior cerebral artery areas. Despite recovering to neurologic baseline, the child was eventually removed from the transplant list secondary to ongoing renal failure, cachexia and pulmonary disease. After extensive consultation with the family, the palliative care team and the ethics committee, a decision was made to discontinue VAD support and allow natural death to take place. Out of concern that the patient would experience unnecessary anxiety, the patient was not informed of the plan, an approach endorsed by the family and both the palliative care and ethics teams. The boy was given an appropriate sedative dose and died peacefully shortly after the VAD was discontinued.

### Clinician perspectives on CD

Requests to discontinue VAD support are not uncommon in adults.<sup>17</sup> Despite this, recent survey studies demonstrate that both patients and caregivers possess a limited understanding of the legal and ethical issues surrounding CD.<sup>18</sup> In both North America and Europe, nearly one third of clinicians continue to view VAD support as different from other life-sustaining measures (e.g., mechanical ventilation, dialysis or inotropes), and demonstrate wide variability in their comfort with CD. In one study, 17% of clinicians responded that they had refused to deactivate a VAD at least once.<sup>18</sup> Various perceptions regarding CD abound, with many clinicians viewing CD as a form of euthanasia, and even more believing that CD is only appropriate in patients who are dying or that discontinuing VAD support is not appropriate for patients admitted for hospice care.

### Ethical issues

CD of VAD support is considered to be both legal and ethical, and is not considered a form of euthanasia because no new pathology is introduced, and the patient dies from the natural progression of his/her underlying disease.<sup>19–21</sup> Regardless of whether or not a patient is actively dying, the underlying principle of patient autonomy dictates that no patient should be obligated to continue a therapy if he/she considers the burden of therapy to outweigh its benefits. Furthermore, choosing to discontinue a life-sustaining therapy is considered ethically indistinguishable from choosing not to initiate the therapy.<sup>13,14</sup> Finally, discontinuation of VAD support is not considered a requisite for inpatient hospice care.

### Consensus guidelines, preparedness planning and deactivation checklists

Guidance with regard to CD in adults has been documented in the literature and supported by professional societies.<sup>17,22</sup> Both the Heart Rhythm Society of America and the European Heart Rhythm Association have published guidelines articulating the ethical principles surrounding deactivation of implantable cardiac defibrillator (ICD) therapy, crafting them to extend beyond just that of ICDs alone by noting that the personal right to refuse or withdraw a treatment “does not depend on the type of the treatment.”<sup>13</sup> Although the International Society for Heart and Lung Transplantation has no formal position regarding CD, their most recent guidelines regarding mechanical circulatory support recommend that palliative care consultation be strongly considered in all patients before VAD implantation, particularly those receiving DT.<sup>23</sup>

In recent years several useful documents have emerged to guide patients and caregivers through palliative care and CD decision-making. Swetz et al have outlined recommendations for a preparedness planning document, a disease-specific and more detailed version of the advanced directive, suggesting that one be completed in all patients before VAD implantation.<sup>16,24</sup> The purpose of this document is to clarify the patient’s wishes not only following a catastrophic complication, but also in the event of device failure, the development of debilitating co-morbid conditions and/or degradation in quality of life. Schaefer et al published an interdisciplinary checklist for VAD deactivation to ensure the least traumatic death experience.<sup>15</sup> However, none of these guidelines specifically address CD of VAD in pediatric patients.

### Withdrawal of life-sustaining therapies in pediatrics

Withdrawal of life-sustaining therapies in pediatric patients is different than that of adults in many ways. First, decision-making by altruistic surrogates, most commonly parents, is the norm. Second, issues of informed consent and patient assent are complicated by variability in age-related developmental and cognitive status as well as severity of illness, which may compromise communication. Uncertainty in prognosis, including the unique capacity for recovery, growth and developmental progress in children after major medical setbacks (e.g., in the case of functional status after a stroke in a toddler), remain ongoing challenges for pediatric providers.

The field of pediatric palliative care is well established, with foundations in neonatology, pediatric oncology and intensive care. Explorations of ethical and legal issues, as well as guidance regarding withdrawal of life-sustaining therapies in children, have been published in consensus statements.<sup>25–30</sup> The application of palliative care concepts in pediatric advanced heart failure, however, is relatively new, as the field of pediatric cardiology has been primarily focused on interventions aimed at repair of defects and prolonging life. With increased use of mechanical support in

this population, issues surrounding CD in particular are of increasing importance. Morell et al recently described the patterns of care for a cohort of 111 children with advanced heart disease who died in hospital. In the 24 hours preceding death, half of the patients were receiving mechanical circulatory support (extracorporeal membrane oxygenation or VAD), with 76 patients (68%) having had discontinuation of disease-directed interventions by the treatment team.<sup>31</sup> In a larger study of end-of-life practices from the Collaborative Pediatric Critical Care Research Network, 9% of children had mechanical support withdrawn, illustrating that CD is an emerging part of the current care paradigm for critically ill children.<sup>32</sup>

### Need for pediatric guidelines specific to withdrawal of VAD support

Because VAD support and DT in particular are much less common in children than adults, fewer pediatric providers have adequate experience with CD. Despite the existence of universal concepts with regard to terminating life-sustaining therapies in pediatrics, a unique psychology accompanies the removal of each form of life-sustaining care. This includes individual variability in the medical provider's capacity to cope with their patient's death, which may be affected by the perception that CD involves a more "active" role in hastening their patient's death than with the withdrawal of other forms of life-sustaining care. For example, patients, caregivers and clinicians may have different feelings associated with discontinuing ventilator support versus stopping artificial nutrition or hydration. Similarly, deactivation of VAD support in a child will also be informed by its own unique set of legal, ethical and cultural issues that must be addressed, despite significant overlap with other forms of end-of-life care. Although inquiries into parent and provider perceptions of prognosis and end-of-life experiences in children with cardiac disease have begun, there is a pressing need for studies to shed light on clinician, family and child attitudes regarding compassionate withdrawal of VAD support as well as consensus guidelines from professional societies.<sup>33,34</sup>

In conclusion, as VAD support for pediatric patients increases in frequency, the pediatric heart failure and palliative care communities must work toward establishing guidelines to clarify the complex issues surrounding CD. Patient, family and clinician attitudes must be ascertained and education regarding the psychological, legal and ethical issues should be provided. Furthermore, pediatric-specific planning documents for use before VAD implantation as well as deactivation checklists should be developed to assist with decision-making at critical points during the illness trajectory. In the interim, we recommend that individual providers considering children for VAD support initiate conversations with patients and their surrogates regarding end-of-life wishes, including the possibility of CD, before device implantation. Moreover, we recommend that programs currently offering VAD implantation in children begin to develop internal policies with regard to CD in conjunction with their institutional palliative care team,

ethics committee and interdisciplinary care providers before the need for CD arises.

### Disclosure statement

The authors have no conflicts of interest to disclose.

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