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Response

To the Editor:

Rady and colleagues1 have raised concerns about the implementation of our guidelines2 and clinicians’ intentions in end-of-life care. We acknowledge that our guidelines do not eliminate moral confusion about end-of-life care in the ICU, and we agree that a focus on intention should be included in any moral appraisal of human action. While our guidelines, template physician note, and order set do not regulate the integrity of clinicians’ actions, they do provide a normative basis around which consensus regarding best practices can develop. That is, they provide a standard framework for developing and communicating the goals of care and support consistency in efforts to achieve them.

Guidelines for the withdrawal of life-sustaining treatment and other palliative measures of last resort, such as palliative sedation, make a distinction between palliation and euthanasia not only on the basis of clinician intention (symptom relief vs patient death), but also on the basis of methods (use of sedative medications sufficient to relieve symptoms vs administration of lethal medications) and the definition of successful outcomes (removal of treatments that are no longer desired or do not provide comfort vs patient death).3,4 Moreover, the rationale for permitting patients and their surrogate decision makers to stop life support is based not only on clinician intention, but also on patient autonomy and informed consent, and the principle of proportionality.4,5 Miller and Truog6 have observed that this rationale exists apart from the question of whether withdrawing life support causes death. The principle of proportionality requires us to consider the patient’s condition (eg, intensity of suffering, expected survival), the anticipated benefits of withdrawal of life-sustaining treatment (minimization of suffering), and the expected harms (possible shortening of survival time); then, to conclude that the cessation of life-sustaining treatment is the most proportional action among the available choices.4

Rady and colleagues1 expressed concern regarding the titration of medications to achieve comfort. Titration is central to successful symptom management in palliative medicine. Moreover, opioids and sedatives may prolong life rather than hasten death after ventilator withdrawal in critically ill patients.6

In delivering palliative care at the end of life, the subjective experience of patients and family is of paramount importance; objective metrics are typically less relevant. The decision made by the patient or a surrogate decision maker to shift treatment goals to comfort care is consequential: the primary goal of health-care providers shifts to keeping the patient comfortable. Except for the magnitude and the immediacy of the consequences, such a choice is like others we support in the course of clinical care. Accordingly, the withdrawal of life-sustaining treatment in these situations honours the individual’s autonomy and is the most proportional response in desperate circumstances without a more desirable and achievable outcome. Finally, it continues to be our impression that our guidelines, template physician note, and order set support patient autonomy at the end of life, as well as strengthen understanding about palliative care practices in our ICU, reduce ethical conflicts, and improve patient care.

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The authors have no conflicts of interest to disclose.

Autosomal-Dominant Polycystic Kidney Disease

To the Editor:

I read with interest the article in a recent issue of CHEST (January 2009) by Li and colleagues.1 However, some points need to be clarified. First, the abbreviation ADPKD stands for autosomal-dominant polycystic kidney disease. Second, the authors think of ADPKD as being unlikely based on the absence of a family history...
and by the presence of pulmonary cysts. While a spontaneous mutation occurs in 5% of cases, one fourth of patients with newly diagnosed ADPKD report no family history. It is noteworthy that a case of numerous pulmonary cysts was reported in a patient with ADPKD, around the time of the first submission of this article.

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Response

To the Editor:

We appreciate the comments of Dr. Kittisupamongkol on our recent report in CHEST (January 2009) on a woman with multiple cysts in the lungs and kidneys that we suspected were due to the TSC2-PKD1 contiguous gene syndrome. As Kittisupamongkol points out, we inadvertently referred to the abbreviation “ADPKD” as “adult dominant polycystic kidney disease” instead of “autosomal-dominant polycystic kidney disease,” and we apologize for the oversight. Kittisupamongkol mentions that the absence of a family history of renal cysts should not preclude a diagnosis of ADPKD and, by citing a recent case report, implies that our patient’s findings might all be due to ADPKD. Although ADPKD is a systemic disease that can involve multiple organs, pulmonary cysts are exceedingly uncommon, to the degree that they are rarely mentioned as an extrarenal manifestation of ADPKD. In an individual with a family history of ADPKD, radiographic evidence of bilateral, fluid-filled renal cysts establishes the diagnosis of ADPKD.

However, we would argue that the diagnosis of ADPKD cannot be made confidently in the absence of a positive family history when extrarenal findings rarely associated with ADPKD (e.g., diffuse pulmonary cysts) and potentially representing a manifestation of another disease (e.g., lymphangioleiomyomatosis or tuberous sclerosis complex) are present. Given the apparent extreme rarity of lung cysts in patients with ADPKD, we believe the more likely diagnosis in our patient is the TSC2-PKD1 contiguous gene syndrome. We wonder whether the individual described by Shanmuganathan and colleagues in their case report and other patients with kidney and lung cysts might also have TSC2-PKD1 contiguous gene syndrome. In any case, definitive diagnosis requires molecular genetic testing (e.g., linkage analysis and/or direct DNA sequence analysis), which was declined by our patient and which was not reported by Shanmuganathan and colleagues.

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