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April 25, 2019

## Dear EMS Organization:

CARES Foundation, Inc. is a non-profit organization that provides support to individuals and families affected by Congenital Adrenal Hyperplasia (CAH). Individuals with adrenal insufficiency, such as CAH, Addison Disease, or other conditions for which glucocorticoids are prescribed, are frequently at risk for fatal adrenal crisis during illness or injury. Increasingly, EMS authorities are recognizing the need for the immediate administration of patient-carried or ambulance-stock injectable hydrocortisone for the treatment of suspected adrenal crisis (NASEMSO, 2018; NAEMSP, 2016).

With up to 3% of the US and UK populations currently taking adrenal-suppressing glucocorticoids (Woods, 2015), there is a substantial portion of the population who lack the ability to mount an adequate cortisol response during times of physical stress. Prompt intervention to prevent an adrenal crisis can significantly reduce mortality (up to 50%) (Willis, 2014).

Since 2009, CARES Foundation has advocated nationwide for local and state EMS agencies to adopt adrenal insufficiency protocols permitting the administration of injectible hydrocortisone, the addition of injectable hydrocortisone to formularies, and training to recognize the signs of adrenal crisis. The seminal American College of Emergency Physicians (ACEP) article

[ <a href="https://webapps.acep.org/CriticalDecisionsTesting/PDFpubs/2012-07-july.pdf">https://webapps.acep.org/CriticalDecisionsTesting/PDFpubs/2012-07-july.pdf</a> ] states the following: "Hydrocortisone is the steroid of choice in an acutely ill patient presenting with acute adrenal crisis because it provides both glucocorticoid and mineralocorticoid effects." A number of states now address acute adrenal insufficiency through EMS protocols and training; <a href="yet-the-re-is-a lack of consistency in medication preference and dosing guidelines">yet, there is a lack of consistency in medication preference and dosing guidelines across the US.

This EMS advisory reflects evidence-based treatment for adrenal crisis (Endocrine Society consensus-based guidelines, 2018). Several supporting organizations, whose community is affected by the risk of death due to adrenal crisis, are endorsing this advisory. Adherence to these recommendations will minimize subsequent need for medical intervention and decrease unnecessary draws on medical resources.

The National EMS Information System (NEMSIS) has included a medical adrenal insufficiency code (9914195<sup>i</sup>) to ALERT EMS staff of this lifethreatening situation. It is catalogued/classified at the NEMSIS database alongside 9914197 (Apparent Life-Threatening Event-ALTE).

The ED (emergency department) code for "Addisonian"/adrenal crisis is listed at the URL: <a href="https://www.acep.org/administration/reimbursement/ed-facility-level-coding-guidelines/">https://www.acep.org/administration/reimbursement/ed-facility-level-coding-guidelines/</a> Under CPT 99291, "there is a high probability of imminent or life-threatening deterioration in the patient's condition" (accessed 1/2/2018). This "CRITICAL CARE" code designates a "crisis state" as time-sensitive.

EMS officials and physicians, in particular, may draw their attention to the following considerations:

- 1) Symptoms (e.g., nausea, vomiting, hypotension, altered mental status, etc.) may not always be present in suspected or impending/imminent/anticipated adrenal crisis. Adrenally-insufficient patients may be identified by medical-alert jewelry, medical-alert card (weight-based and/or age-based instructions), or medical letter. Presentation of a vial of patient-held injectable hydrocortisone incites verification of adrenal insufficiency status and protocol adherence. Anticipated symptoms in adrenal crisis should NOT be used as a prerequisite to institute treatment.
- 2) If in doubt, medical control should be contacted to approve administration of injectable hydrocortisone in the field. Patient outcome is potentially fatal if the injection is delayed; however, prompt administration in the pre-hospital setting could prevent/treat adrenal crisis and avert severe morbidity or mortality. Patients should be transported to the closest emergency department and adrenal insufficiency/adrenal crisis status communicated.
- 3) There is no clinical rationale to restrict/prohibit injectable hydrocortisone for adrenal crisis to a particular age group (*e.g.*, no contraindication in newborns or children under 1 year of age)
- 4) Preservatives (benzyl alcohol) in injectable hydrocortisone formulations should not deter/delay paramedics from administering medication to infants. Medical control should be contacted, and the emergency department should be alerted upon arrival regarding the risks associated with benzyl alcohol in newborns.
- 5) Other pre-hospital protocols should be considered as appropriate to treat hypotension, hypoglycemia, hypovolemia, hyponatremia, shock, anaphylaxis, drug overdose, sepsis, bleeding, seizures, *etc.*
- 6) If budget permits, we recommend the stocking of injectable hydrocortisone on ambulances and the use of EMS on-line training modules if existing e-learning systems are in place. (e.g., <a href="https://hscssl.unm.edu/EM/PED/emsc/training/adrenal/adrenal.html">https://hscssl.unm.edu/EM/PED/emsc/training/adrenal/adrenal.html</a>)

This EMS advisory clarifies the immediate needs of a person in adrenal crisis. No longer will EMS personnel have doubts regarding the immediate administration of the right medication at the right dose to save the life of someone with adrenal insufficiency.

Thank you for your assistance in sharing this information with your membership. Please do not hesitate to contact us with feedback or questions.

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Sincerely,

Dina M. Matos Executive Director Karen Lin Su, M.D. Medical Director

ihttp://www.nemsis.org/media/nemsis\_v3/3.4.0.150302/DataDictionary/APIs/EMSFrameDoc/schemas/commonTypes\_v3\_xsd/simpleTypes/ProtocolsUsed.html (accessed 7/11/2018)