Palliative Sedation with Oral Medicines in a Neonate with Herlitz-Type Epidermolysis Bullosa (FR419-A)

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Objectives

- Identify two challenges of providing palliative sedation via the enteral route.
- Identify at least two sedative medications that are typically administered via intravenous route but are safe for oral/buccal administration in a patient who has no IV access.

Background: Palliative sedation (PS) is commonly used for management of refractory symptoms at or near end of life, most often with medication administered via a combination or enteral and parenteral routes. We present the case of a neonatal male diagnosed with junctional epidermolysis bullosa managed exclusively with oral analgesic/sedative agents, including the use of oral agents after palliative sedation was initiated.

Case Description: Junctional epidermolysis bullosa (JEB) is a devastating disease characterized by mucocutaneous blistering that can present at birth, often leading to early mortality. Infants with the severe form of JEB typically die by 12 months of age. For our patient, with early goals of care discussions, the family opted for aggressive comfort care, including a request that no invasive lines, tubes, or drains be placed. Our pediatric palliative care team managed his pain using conventional oral analgesic agents including acetaminophen, opioids, benzodiazepines, gabapentin, and oral ketamine. This combination and escalation of doses was sufficient for approximately 4.5 months. At age 5 months, the patient developed pain and respiratory distress for which he was no longer able to be awake and comfortable despite multiple medication changes and dose increases, so PS was initiated. This was a challenge given the inability to administer medications via intravenous or subcutaneous routes. Our patient received a combination of oral and buccal pentobarbital, diphenhydramine and fentanyl with immediate relief of suffering while remaining in a state of moderate to deep sedation. Although ordered, dexmedetomine was never used. The patient died 5 days after initiation of the PS and was peaceful throughout.

Conclusion: Although not a conventional means by which to administer PS, oral and nasal medicines can be used effectively and safely to provide relief of refractory symptoms when IV access is not feasible.