The view from up here

Treatment of acute sickle cell pain in pediatrics: An ethical analysis from a high-altitude medical school

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Introduction

- Sickle cell disease (SCD) is a lifelong condition characterized by acute pain crises that overwhelmingly affects POC
- Contextualization of the interfaces between SCD patients and the healthcare system early in life will aid in trending, characterizing, and ultimately improving those interactions
- Objective: to assess the current state of acute SCD pain treatment in pediatrics through the lens of bioethics and to understand gaps in education for medical students who have limited contact with SCD patients

Epidemiology

- 100,000 people in the US are living with SCD
- 1 in 365 Black live births (1 in 13 have sickle cell trait)
- 1 in 16,300 Latinx births
- Only 16-87% of cases captured through data collection program over 14 years in two states
- Newborn blood screening programs have existed for 50 years but have not become standardized nationally
- 95% of all infants born in the US with SCD are expected to survive to adulthood
- SCD patients have a 20-year reduction in lifespan globally and a mean life expectancy of 43 years in the US

High Altitude

- Identification and avoidance of known sickling triggers is a pillar of SCD management
- A 1981 report recommended that all SCD patients without previous exposure to mountain environments avoid the mountains
- Current recommendations include preparing for travel to altitude early with gradual conditioning (when possible)
- Living at lower altitude is likely due to a combination of factors: past recommendations, access to care, family and community
- High-altitude providers and hospital systems expect to see fewer SCD patients in the clinical setting

Pathophysiology

- SCD is an autosomal recessive inherited hemoglobinopathy
- A single nucleotide substitution results in hemoglobin polymerization → red blood cells sickle and hemolyze
- The resultant vaso-occlusion leads to acute episodes of severe pain (called vaso-occlusive crisis, or VOC)
- Vaso-occlusion can also cause progressive multiorgan damage and increased mortality
- The mainstay of treatment for VOC pain is opioid therapy
- More research is being done to further characterize VOC pathogenesis to better target therapies

Pediatric SCD

- Mortality has fallen since the introduction of a vaccine against pneumococcal disease in 2000
- The transition to adult care is particularly challenging and is associated with higher mortality
- Health-related quality of life (HRQOL) and emergency department reliance (EDr) are significant areas of research → data indicate that modifiable risk factors in SCD morbidity and mortality are lifelong and intertwined with medical, social, economic, and political factors

Application & Future Directions

- Multifactorial care gaps result in undertreated VOC pain
- Medical students in Colorado will have less exposure to SCD than their peers at lower elevations
- SCD is currently taught through genetics, hematology, and infectious disease
- A parallel ethics and humanities curriculum could integrate the social determinants of health and the barriers to care for SCD patients
- Increased curricular exposure and comfort with SCD and VOC will contribute to higher quality patient care for this population as students transition to residency in other regions of the US

The Four Tenets of Bioethics:

Beneficence: acting in the benefit of the patient

- SCD patients often require higher doses of opioid medication secondary to tolerance after repeated and regular exposure
- Many SCD patients are labeled as "drug-seekers" which invites biases against this population among medical providers
- The righting reflex inherent in medicine may manifest as relying entirely on opioid therapy for VOC; however, many patients benefit from a longitudinal and multidisciplinary approach

Justice: fairness and equity

- The vast majority of SCD patients are POC: issues of justice are amplified in this patient population
- Placing patients experiencing VOC in a triage order in the emergency department is especially challenging
- The establishment of SCD-specific pain protocols have been shown to be effective

Nonmaleficence: the avoidance of harm or injury

- The bare minimum for the standard of care in the medical profession
- Providers must consider the sequelae of their treatment plans: patients who receive large doses of opioid medication are at risk for respiratory compromise and must be closely monitored
- Taking a multidisciplinary approach to VOC pain in the acute setting takes more time and resources but can result in better long-term outcomes

Patient Autonomy: patient decision-making in an informed and voluntary manner

- "Non-compliance" as a label for patients who do not follow all medical recommendations (regardless of the reason) was born out of paternalistic medicine and results in an antagonistic patient-provider relationship
- Time is the limiting factor for the ED provider treating VOC → providers must rely on heuristics and written policies
- By entering the hospital system, SCD patients relinquish autonomy in exchange for pain management

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