

Implementation of a Caregiver-Focused Educational Intervention to Improve Knowledge and Competency for Managing Adrenal Crisis in Patients with Duchenne Muscular Dystrophy

Mirini Kim, MS, CPNP-PC

Brigit VanGraafeiland, DNP, CPNP, FAAN

Rachel I. Gafni, MD

Objectives

- Discuss background and significance of adrenal crisis (AC) education and management in Duchenne Muscular Dystrophy (DMD)
- Discuss project purpose, aims, and methods
- Review project results and implications.

Introduction

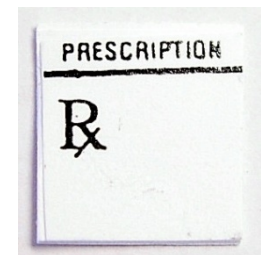
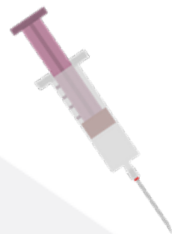
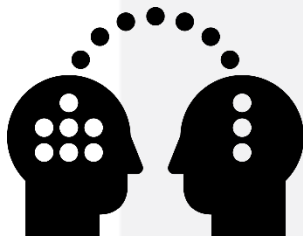
- Risk of AC in DMD patients
- Untreated AC can lead to hypoglycemia, hypotension, cardiovascular collapse, and death
- Paucity of literature in DMD population

Background of the Clinical Problem

- The 2018 DMD Care Considerations
 - Specific AC-related recommendations
- Baseline data in clinic
- Implementation requires change in practice
 - Caregiver
 - Providers

Project Aims

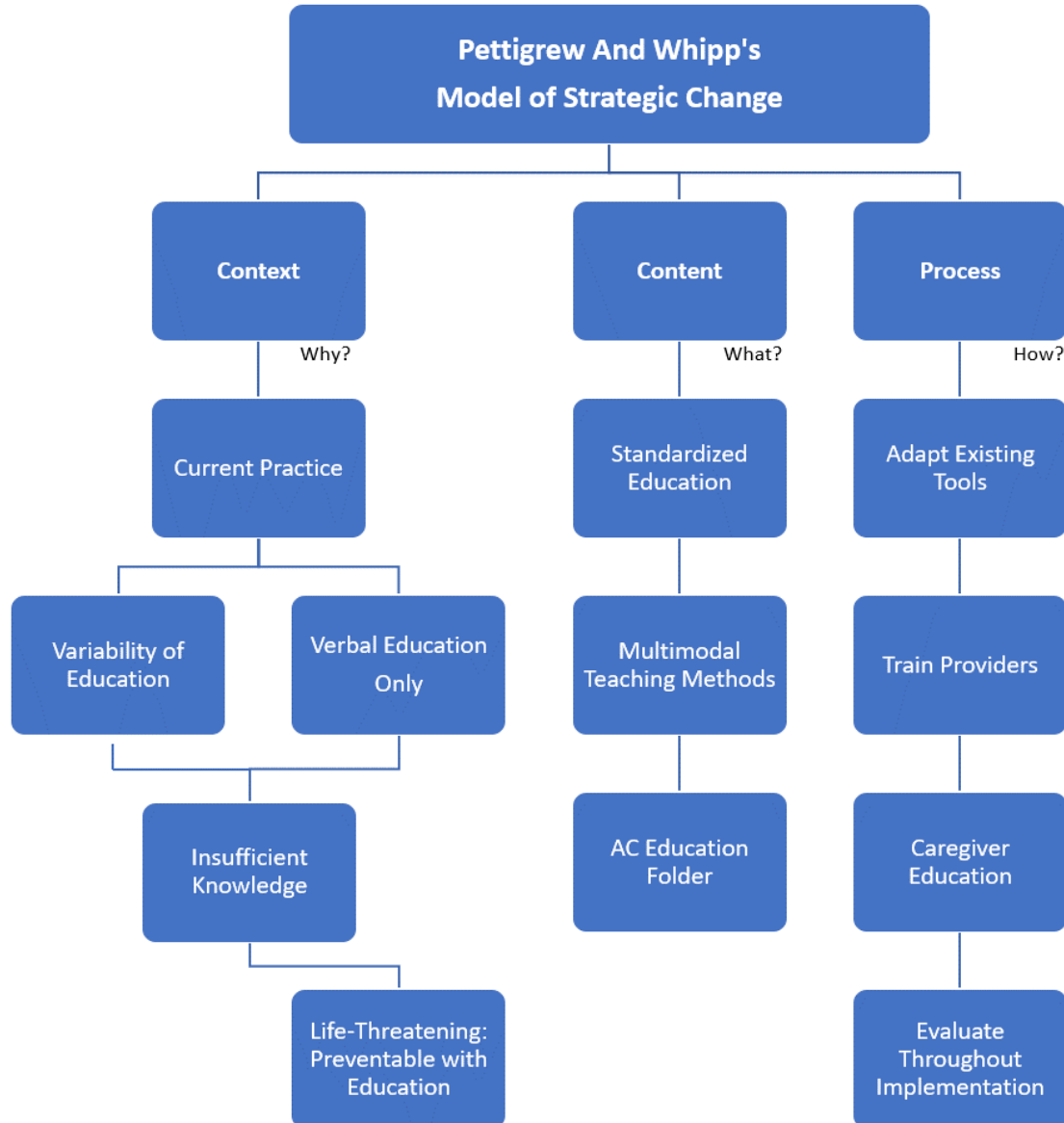
- 1) Evaluate if a caregiver-focused educational intervention improves knowledge of AC management
- 2) Determine the number of iterations required for caregivers to demonstrate injection competency
- 3) Determine if there is an increase in prescription rates of emergency hydrocortisone following provider training



Review of the Literature

- Despite prior education, patients had inadequate understanding of preventing AC
- Inadequate knowledge regarding how to administer emergency hydrocortisone injection
- Verbal education alone is insufficient
- Effectiveness of teach-back method

Translation Framework



Methods

- **Design:** Quality improvement pre-post study
- **Sample:** caregivers of patients with DMD and neurology providers at an academic medical center in the mid-Atlantic

Methods

- **Intervention:** Education sessions with each caregiver and provider
 - **Caregivers:** AC and Injection training
 - **Providers:** PowerPoint presentation and quick reference
- **Data Collection**
 - Knowledge questionnaire pre and post caregiver education
 - Injection Competency: Checklist
 - Rate of prescriptions by providers via chart review

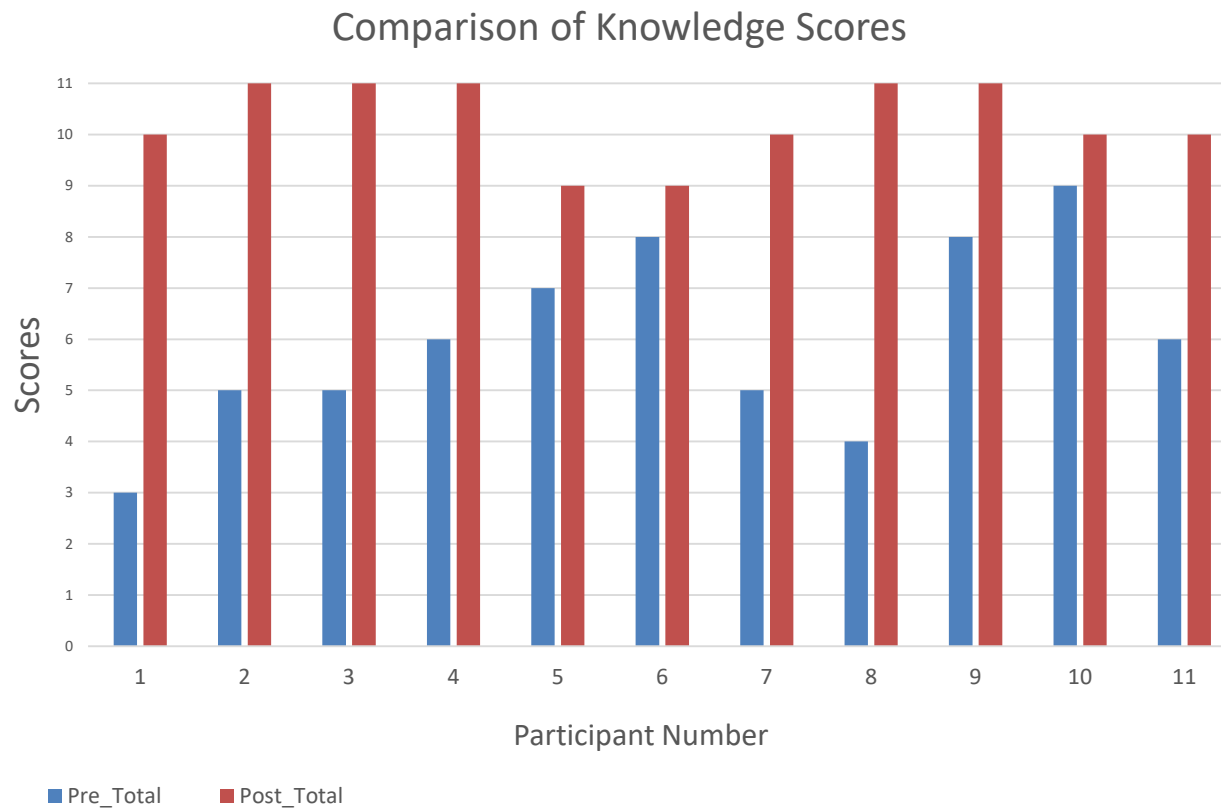
Results

- Caregiver Demographics

Caregiver Demographics	(N=11)
Age, n (%)	
21 - 30 years old	2 (18.2)
31 - 40 years old	6 (54.5)
41 - 50 years old	3 (27.3)
Sex, n (%)	
Male	4 (36.4)
Female	7 (63.6)
Relationship to patient, n (%)	
Parent	10 (90.9)
Legal Guardian	1 (9.1)
Highest level of education, n (%)	
High school diploma/GED	3 (27.3)
Associate's degree	3 (27.3)
Bachelor's degree	3 (27.3)
Master's degree	2 (18.2)

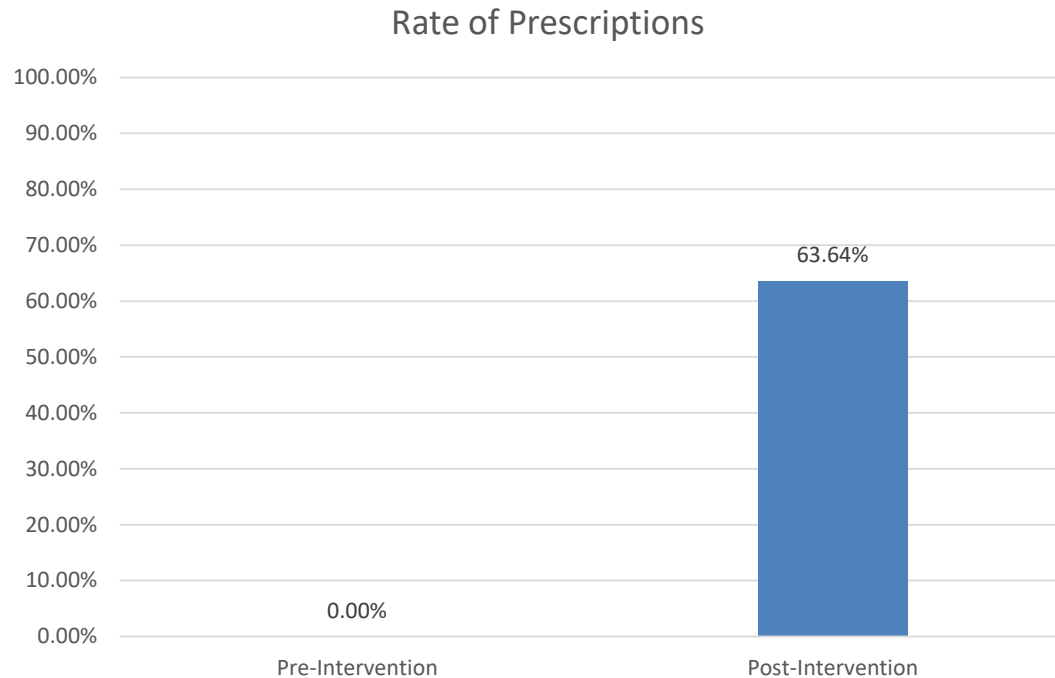
Results

- Aim 1



Results

- Aim 2: All caregivers demonstrated injection competency in one iteration
- Aim 3:



Discussion

- Results were consistent with the existing literature
 - Multimodal education improves knowledge and competency
- Provider presentation and a visual reminder in the clinic space supported increase in emergency Rx rate
- Strengths and Limitations

Conclusions

- Swift implementation will protect from the life-threatening complications of AC
- Future Research
 - A standardized education for DMD
 - Studies that compare the effects of different teaching approaches
 - Frequency of education
 - Barriers to implementation

Translation into Practice

- Dissemination
 - Nursing leadership
 - The Parent Project Muscular Dystrophy
 - Greater DMD community
- Sustainability
 - AC champion in the clinic
 - Share AC education resources to department

References

- Antje, N., Stefan, J., & Emanuel, C. (2018). Evaluation of the frequency of adrenal crises and preventive measures in patients with primary and secondary adrenal insufficiency in Switzerland. *Swiss Medical Weekly*, 148(3-4) doi:10.4414/smw.2018.14586
- Arcebido, R., Wong, E., Cohen, V., Likourezos, A. (2013). Pharmacist-led discharge counseling on subcutaneous insulin use and administration. *American Journal of Health-System Pharmacy*, 70 (16), 1371-3. doi: 10.2146/ajhp120571
- Braatvedt, G. D., Newrick, P. G., & Corrall, R. J. (1990). Patients' self administration of hydrocortisone. *BMJ (Clinical Research Ed.)*, 301(6764), 1312.
- Birnkrant, D. J., Bushby, K., Bann, C. M., Alman, B. A., Apkon, S. D., Blackwell, A.,...Ward, LM (2018). Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and neuromuscular, rehabilitation, endocrine, and gastrointestinal and nutritional management. *Lancet Neurology*, 17 (3), 347-361. doi: 10.1016/S1474-4422(18)30024-3
- Correnti, C., Cassidy, L., Chen, S. C., & Stoff, B. (2017). Video patient education about systemic corticosteroids is more effective than verbal education: A randomized controlled trial. *Journal of Investigative Dermatology*, 134, S95. doi:10.1038/jid.2014.109
- Dantic, D.E. (2014). A critical review of the effectiveness of “teach-back” technique in teaching COPD patients self-management using respiratory inhalers. *Health Education Journal*, 73(1), 41-50. doi: 10.1177/0017896912469575
- Department of Endocrinology at Children’s National Medical Center. Solu-Cortef injection education and training checklist. Children’s National Health System, Washington, DC.
- Fleming, L. K., Rapp, C. G., & Sloane, R. (2011). Caregiver knowledge and self-confidence of stress dosing of hydrocortisone in children with congenital adrenal hyperplasia. *Journal of Pediatric Nursing*, 26(6), 55. doi:10.1016/j.pedn.2011.03.009
- Flemming, T. G., & Kristensen, L. O. (1999). Quality of self-care in patients on replacement therapy with hydrocortisone. *Journal of Internal Medicine*, 246(5), 497-501. doi: 10.1046/j.1365-2796.1999.00538.x
- Harsch, I. A., Schuller, A., Hahn, E. G., & Hensen, J. (2010). Cortisone replacement therapy in endocrine disorders - quality of self-care. *Journal of Evaluation in Clinical Practice*, 16(3), 492-498. doi:10.1111/j.1365-2753.2009.01149.x
- Kampmeyer, D., Haas, C. S., Moenig, H., & Harbeck, B. (2017). Self-management in adrenal insufficiency - towards a better understanding. *Endocrine Journal*, 64(4), 379-385. doi:10.1507/endocrj.EJ16-0429
- King, J., Mitchelhill, I., Fischer, M.J. (2007). Development of the congenital adrenal hyperplasia knowledge assessment questionnaire (CAHKAQ). *Journal of Clinical Nursing*, 17(13), 1689-1696. doi: 10.1111/j.1365-2702.2006.01851.x

References

- Kinnett, K. & Noritz, G. (2017). The PJ Nicholoff steroid protocol for Duchenne and Becker Muscular Dystrophy and adrenal suppression. *PLoS Currents*, 9. Retrieved from <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5505768/>
- McDonald, C. M., Henricson, E. K., Abresch, R. T., Duong, T., Joyce, N. C., Hu, F.,...Gordish-Dressman, H. (2018). Long-term effects of glucocorticoids on function, quality of life, and survival in patients with Duchenne muscular dystrophy: a prospective cohort study. *Lancet*, 391, 451-461. doi:10.1016/S0140-6736(17)32160-8
- National Quality Forum (2010). Safe practices for better healthcare - 2010 Updates: A consensus report. Retrieved from https://www.qualityforum.org/Publications/2010/04/Safe_Practices_for_Better_Healthcare_%E2%80%93_2010_Update.aspx
- Puar, T., Stikkelbroeck, N., Smans, L., Zelissen, P., Hermus, RM (2016). Adrenal crisis: Still a deadly event in the 21st century. *The American Journal of Medicine*, 129 (3), 339.e1-9. doi: 10.1016/j.amjmed.2015.08.021
- Repping-Wuts, H. J., Stikkelbroeck, N. M., Noordzij, A., Kerstens, M., & Hermus, A. R. (2013). A glucocorticoid education group meeting: An effective strategy for improving self-management to prevent adrenal crisis. *European Journal of Endocrinology*, 169(1), 17-22. doi:10.1530/EJE-12-1094
- Shulman, D.I., Palmert, M. R., Kemp, S. F. (2007). Adrenal insufficiency: Still a cause of morbidity and Death in Childhood. *Pediatrics*, 119 (2), e484-94. doi: 10.1542/peds.2006-161
- South Australian Health (2013). Guide for engaging with consumers and the community 2013. *The Government of South Australia*. Retrieved from https://www.sahealth.sa.gov.au/wps/wcm/connect/f8d1d0004e454788aa0caf8ba24f3db9/Guideline_Guide+for+Engaging+with+Consumers+and+Community_Oct2015.pdf?MOD=AJPERES&CACHEID=f8d1d0004e454788aa0caf8ba24f3db9
- Stetler, C. B., Ritchie, J., Rycroft-Malone, J., Schultz, A., & Charns, M. (2007). Improving quality of care through routine, successful implementation of evidence-based practice at the bedside: An organizational case study protocol using the Pettigrew and Whipp model of strategic change. *Implementation Science* 2(3).
- Truong, T.H., Nguyen, T.T., Armor, B.L., Farley, J.R. (2017). Errors in the administration technique of insulin pen devices: A results of insufficient education. *Diabetes Therapy*, 8, 221-226. doi: 10.1007/s13300-017-0242-y
- van der Meij, E. J., Gobbens, R. J., Beukers, J., Geilvoet, W., van der, L. A., & Neggers, S. J. (2016). Much to be desired in self-management of patients with adrenal insufficiency. *Int J Nurs Pract*, 22(1), 61-69. doi:10.1111/ijn.12368

Thank you!

- Dr. VanGraafeiland and Dr. Rachel Gafni
- Dr. Laura Tosi (Ortho and Bone Health)
- Dr. Andrea Estrada (Endocrinology)
- Children's National Hospital Neuromuscular Team

