Using behavioral skills training to teach adolescents with developmental disabilities interview skills
Tracie Mann, Ph.D., LBA and Kimberly James-Kelly, M.A., LBA
Child Study Center

Abstract

Background: Adolescents and adults with disabilities are underemployed compared to their peers without disabilities. They have more difficulty navigating all components of employment: finding job opportunities, successfully interviewing for a job, and maintaining a job once hired. Teaching adolescents and young adults with developmental disabilities interview skills is an important first step toward securing employment. There are some data to suggest that Behavioral Skills Training is effective in this endeavor, but more are needed. The purpose of this study was to evaluate the effects of Behavioral Skills Training on interview skills for two adolescents with developmental disabilities.

Methods: We used a multiple-probe design to measure the effects of Behavioral Skills Training on the interview skills of two adolescents. In baseline, we administered an interview to both participants without any instruction on how to answer the questions. The interviews consisted of 23 questions grouped into four different categories: Personal Information, Experience, Logistics, and Summary questions. We measured how they responded both verbally (the content of the answers) and non-verbally (eye contact, posture, tone, and volume) to each question. Following baseline, we began using Behavioral Skills Training to teach appropriate verbal and non-verbal responding to questions in Unit 1. Following instruction in each unit, we administered another full interview until all units had been taught and a final interview administered.

Results: Both participants performed poorly in baseline, each only answering one question correctly and failing to demonstrate appropriate non-verbal behavior consistently. Behavioral Skills Training resulted in improved performance for both participants across all question units. Further, performance only improved following instruction in the targeted unit, thus demonstrating experimental control of the independent variable over responding.

Discussion: The results from the current study are promising. The main limitation is related to the generality of these findings. The two participants may not be representative of the population of adults with developmental disabilities seeking employment. Further, the extent to which the effects of the training will generalize to novel questions or interviewers is unknown. Future research should continue to evaluate the effectiveness of Behavioral Skills Training and address these limitations.

Category: Evidence Based Practice
Minimizing Lung Injury: Everybody Plays a Part
Samantha Corkum, BSN, RNC-NIC, Julie Kirkwood, MS, RRT-NPS, Teri Lynn Bjornlie, RRT-NPS, NICU

Abstract

**Purpose:** Despite our efforts to avoid invasive ventilation, our rate of chronic lung disease (CLD) in the very low birth weight (VLBW) population remained high. In 2016 the rate of CLD in VLBWs had climbed to 37.8%. For patients whom we received immediately after birth, CLD rate was only 15.8%. For patients received after day of life 3, CLD rate was 47.1%. Our goal was to decrease our incidence of CLD to match that of top-performing NICUs in the Vermont Oxford Network (VON), which is 19%, by December 2017.

**Methods/Implementation Strategies:** Our team consisted of multi-disciplinary healthcare workers and parent liaisons. Drivers of Change:

1. Change in surfactant brand and dosing
2. Change in surfactant delivery method (utilize T-piece resuscitator)
3. Increase caffeine dosing
4. Increasing family involvement in Kangaroo Care
5. Utilize family advisors to create a kangaroo care 'how to' video of an infant on a high-frequency jet ventilator (HFJV), so nurses and therapists will be more comfortable transitioning the baby from the bed to the parent.
6. Implementation of a NICU core RRT team

**Results:** Surfactant: From 7/20/16 to 9/26/17 148 infants received the new brand of surfactant, and there was a 30% reduction in the number of infants requiring re-dosing. Family involvement: 100% of NICU bedside staff attended spring training and viewed the kangaroo care educational video. Staff reported that they are more comfortable with and more likely to encourage kangaroo care of an infant on a HFJV. Vent days: We saw a 30% increase in the number of infants never intubated in 2017 vs 2015 and a 37% decrease in those intubated >28 days in 2017 vs 2015. VLBW CLD rate. In 2016 we had an overall CLD rate of 37.8% amongst our VLBWs. For 2017 that rate declined to 26.8%, which is a 29% decrease. For infants discharged as of September 2018, our CLD rates in our VLBW population have continued to decline to 25%.

**Discussion:** While we did not hit our goal of 19%, we saw an impressive reduction in CLD. Looking back, we should have made our goal to be in-line with that of other top-performing regional referral centers within VON, which was 29%. When comparing ourselves to similar facilities (Type C NICUs), our rate of CLD at the end of our project was well-within the top quartile of Type C centers.

Category: Evidence Based Practice
DIFFUSION-WEIGHTED MAGNETIC RESONANCE IMAGING SHOWS A CRITICAL DIFFERENCE BETWEEN SUPRATENTORIAL AND INFRATENTORIAL PEDIATRIC EPENDYMOMA

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Neuro-Oncology, Radiology, and Pathology

Abstract

Background: Magnetic resonance imaging (MRI) features differ between various childhood brain tumors. Advanced MRI techniques including diffusion-weighted imaging (DWI) can add vital information to conventional sequences, potentially crucial in creating an appropriate differential diagnosis. Ependymoma, the third most common malignant brain tumor in youth, remains challenging with respect to an imaging differential diagnosis. We have recognized a critical difference in DWI characteristics between supratentorial (ST) and infratentorial (IT) ependymomas in children.

Methods: Following IRB approval, we re-reviewed the MRIs and neuropathology from 30 consecutively diagnosed children with intracranial ependymomas, 2007-2016, abstracting for tumor location (ST vs IT), diffusion characteristics and tumor grade. Chi-square and Cramer’s V tests were applied.

Results: The mean age at diagnosis was 5 years (18 boys, 12 girls). Eighteen patients had IT tumors and 12 had ST tumors. Ten tumors had anaplastic histology (WHO grade III) and 20 had classic histology (WHO grade II). Eleven tumors showed restricted diffusion on DWI, and 19 did not (13 facilitated diffusion and 6 were equivalent to surrounding brain). All tumors that showed restricted diffusion were ST; none were IT. Of the 11 restricting tumors, 3 had anaplastic histology. Of the 13 non-restricting tumors, 4 had anaplasia. The relationship between tumor location and restricted diffusion was significant and strongly associated ($\chi^2(1) = 26.053$, $p < 0.001$, Cramer’s V= 0.932).

Discussion: Diffusion-weighted imaging can supplement conventional MRI sequences in formulating a critical differential diagnosis in children with brain tumors. Our analysis shows a dramatic difference in diffusion characteristics between ST and IT ependymomas, independent of tumor grade. Supratentorial tumors tend to show restricted diffusion, but IT tumors, irrespective of grade, do not restrict. We postulate that there are physiologic tumor microenvironmental factors that account for this observation. Awareness of DWI differences in ependymoma may be helpful in refining the initial differential diagnosis.

Category: Research Study
Mutation of CEP72 Gene May Predispose Patients to Hepatotoxicity
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Hematology and Oncology Department

Abstract

Background: Polymorphisms in CEP72 have been linked to increased incidence of vincristine-induced toxicities, namely peripheral neuropathy. We hypothesize that polymorphisms in the same gene may also increase a patient’s risk of developing hepatotoxicity when receiving potentially hepatotoxic agents during chemotherapy. This report describes drug-induced cholestasis noted with the use of vincristine and 6-mercaptopurine (6-MP) in a patient homozygous for the CEP72 risk alleles.

Methods: This report employed the use of electronic medical records for the review of this patient's diagnosis, treatment, and outcome at Cook Children's Medical Center.

Results: A 10-year-old female with high-risk B precursor ALL entered remission after induction therapy. She developed acute hepatic injury with elevated direct bilirubin during weeks five and six of consolidation. These weeks exposed her to 6-MP, vincristine, and PEG-asparaginase. We held 6-MP due to its hepatotoxic potential, and reduced the vincristine dose by 75%. Direct bilirubin normalized over time. Cytogenetic testing revealed homozygous CEP72 risk alleles and normal TPMT*1 alleles. During interim maintenance and delayed intensification, the patient received 50% dose reduction of vincristine due to the association between CEP72 mutations and vincristine-induced toxicities. Given normal TPMT status, she received protocol-guided full doses of 6-MP. During maintenance, she proceeded to receive 50% dose reduction of vincristine and full dosing of 6-MP as well as weekly methotrexate. The patient again developed jaundice and elevated direct bilirubin, which normalized following a renewed 50% dose reduction of 6-MP. The patient continues to tolerate therapy well with full doses of vincristine and methotrexate while the dose of 6-MP remains at 50%.

Discussion: Patients who possess polymorphisms in CEP72 may be at higher risk for developing hepatotoxicity when receiving chemotherapy agents for acute leukemia. Animal studies suggest that intact microtubular function enables hepatic uptake of bilirubin. Disruption/inhibition of microtubular polymerization has been shown to interfere with hepatic function. These may form the basis for CEP72 mutations leading to peripheral neuropathy and additionally hepatic injury. In this report we describe a novel adverse effect in association with polymorphism in the promoter region of CEP72. If replicated among other patients, detection of this mutation may allow the treating clinician to dose modify during therapy and improve overall therapeutic outcome.

Category: Case Study
When the Going Gets Tough, Go Psychometric! Evidence-Based Practice (EBP) Strategies towards Ensuring Reliability and Validity of the Pediatric Early Warning System (PEWS)
Jamie Bankston, Mary Cazzell
Nursing

Abstract

Background: An urban pediatric hospital used several PEWS revisions in an effort to reduce cardiac arrests and detect clinical deterioration on inpatient units. Concerns were subjectivity of scored items, non-evidence-based criteria, false positive and negative scores, and tool accuracy. The purpose of this interdisciplinary EBP project was to address PEWS reliability and validity. In pediatric patients, how does an evidence-based PEWS tool compared to non-evidence-based iterations affect consistent measurement of clinical deterioration within a crucial timeframe to prevent delays in intervention?

Methods: A synthesis of reviewed literature revealed gaps including inconsistencies in pediatric vital sign (VS) ranges, as well as objective and non-evidence-based scoring parameters. An opportunity for data collection and analysis was revealed. The Iowa Model aided in structure of the practice question, project team, evidence review, pilot, and evaluation before full implementation A project team reviewed PEWS iterations, revised subjective criteria, developed scoring parameters, and implemented EBP-based pediatric VS ranges. Nurses on a 40-bed medical pilot unit completed education. A modified electronic medical record (EMR) enhanced scoring clarity. For inter-rater reliability data, charge nurses scored patients within 5 minutes of staff nurses.

Results: Physical assessment EMR entries were correlated with cardiovascular and respiratory PEWS categories for convergent validity. With 82 rater dyads, tool reliability was 0.78. Component reliability coefficients were behavior: 0.83; cardiac: 0.67; and respiratory: 0.72. Moderate convergent validity on 17,872 scores was reported as absolute agreement (82%-94%) and kappa (0.48-0.53). Psychometric properties of reliability and validity were established for a revised PEWS.

Discussion: Lack of integration between charted and EMR VS, inconsistency in addition and charting scores were identified barriers. Lessons learned included factors affecting inter-rater reliability. The project provides confident recommendation for house-wide rollout of EBP PEWS, and foundation for further research.

Category: Evidence Based Practice
COMPARING THE USE OF IPAD DISTRACTION VERSUS ORAL VERSED TO REDUCE PEDIATRIC PREOPERATIVE ANXIETY

TERRI PEARCY, RN, BSN, CPN & REBECCA SEVERNAK, RN
Perioperative Services

Abstract

**Background:** Children experience anxiety before outpatient surgical procedures at parental separation and mask induction. Most common treatment for preoperative anxiety is administration of benzodiazepines, usually oral Versed, with undesirable side effects of prolonged sedation and emergence delirium. The research question was: In children (4-12) years undergoing outpatient surgery, is distraction using iPad effective in reducing preoperative anxiety (at parental separation and mask induction) when compared with oral Versed? Purpose was to replicate Seiden et al (2014) study to increase evidence for use of distraction in pediatric perioperative patients.

**Methods:** Single-blinded prospective comparative study design was used with randomized assignment to experimental iPad group (n=51; mean age 6.8 years, 51% male) and control oral Versed group (n=51, mean age 6.9 years, 65% male). Outcome measures included: (1) Modified Yale Preoperative Anxiety Scale at 3 time points, (2) Pediatric Anesthesia Emergence Delirium Scale at 2 time points (3) parent's perception of child's anxiety at 2 time points (4) parent satisfaction with child separation and (5) times from Post-Anesthesia Care Unit (PACU) arrival to awakening and to discharge.

**Results:** iPad distraction was more effective in reducing pediatric preoperative anxiety in pediatric outpatient surgery patients, 4-12 years old, decreasing scores of emergence delirium, and shortened length of stay in PACU.

**Discussion:** Results supported change in practice for perianesthesia nurses related to use of distraction to reduce pediatric preoperative anxiety and decrease overall use of benzodiazepines,

Category: Research Study
Identification of Pediatric Obesity in an Urban Urgent Care Center
Kari L. Guffey
UCC

Abstract

Background: Historically, each generation outlives the last. However, for the first time in recorded history, the current generation of children are predicted to die younger than previous generations because of the pediatric obesity crisis (Cygan, Chehab, Rodriguez & Zenk, 2014). Childhood obesity has increased by 300% in the United States (U.S.) over the past three decades (Radnitz et al., 2013). The consistent increase in pediatric obesity exceeds that of any other pediatric disease. Twenty percent of U.S. children are overweight (Centers for Disease Control, CDC, 2017). Obesity is of epidemic proportions and exposes our children to multiple additional health problems which lead to lifelong consequences (Radnitz et al., 2013). The American Academy of Pediatrics, World Health Organization and US National Institute of Health are recommending that protocols be developed for the management of obese children. Once the children are identified as obese through body mass index (BMI) evaluation, treatment should be targeted and goal-oriented (Radnitz et al., 2013). BMI is the recommended screening tool for child obesity (Krebs, Himes, Jacobson, Nicklas, Guilday & Styne, 2007). Once a BMI is obtained, the CDC recommends plotting each child on the chart with associated percentage recommendations for BMI-To-Age (CDC, 2017).

Methods: For the project, the author will applied the CDC recommendations for identification of overweight and obese children at an urban Urgent Care Center (UCC). Within the continuum of care for pediatric patients, the CDC Growth Charts are the most commonly used indicator to measure the size and growth patterns of children and teens in the United States (CDC, 2017). The author educated the UCC provider staff regarding the breadth and scope of pediatric obesity utilizing educational posters and information sessions. The author disseminated the knowledge current literature shows that clinicians are underreporting the diagnoses of overweight and obese status in the pediatric setting (Thomas & Urrego, 2017). Educational materials were dispersed including appropriate ICD-10 codes for children in the UCC setting who qualify as overweight/obese. A secondary purpose of this data collection was to determine the demographics of the UCC population. This data provided evidence to support a pediatric obesity taskforce within the health network as well as aided in developing weight management protocols geared toward the specific pediatric population in the area.

Results: 672 children ages 4-16 were measured with BMI calculated and populated in the EMR. The author plotted each child on the CDC BMI for age percentile chart and reported the percentile on a data spreadsheet. The median age for the cohort was 8 years old. There was an equal balance between males and females and the ratio of Hispanic to Non-Hispanic children was quite similar. There were 305 (45.4%) of the children who presented to the UCC for care who were overweight/obese. Obesity characteristics of this population included more prevalence in Hispanic versus Non-Hispanic children and a significant number of children who qualified as severely obese.

Discussion: The author recommended that the UCC measure and document height and BMI on all children at all visits. This is a consistently recommended practice and would allow for recognition and education.

Category: Research Study
Addressing Disturbing Trends in Pediatric Suicide: Collaborative EBP Project Utilizing a Suicide Risk Assessment in the Emergency Department
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Psychiatry/Emergency Department

Abstract

Background: From 2015 to 2016, there was a 44% increase in patients with suicide attempts who were admitted to medical units at Cook Children’s Medical Center. Time constraints, stigma regarding mental health, and inadequate training to perform a suicide risk assessment, were identified as barriers for nurses in the ED.

Methods: The Behavioral Health Department and ED collaborated to identify, educate, and implement the evidence-based ASQ suicide screening tool for all patients 10 and over. Pre-Survey was distributed electronically to all ED nurses. Based on pre-survey results, education interventions included: (1) Computer Based Training with audio explaining the benefits of suicide risk screening in the ED; (2) video presentation on the administration of the ASQ; and (3) handout guiding documentation. Post-Survey was distributed 2 weeks after ASQ implementation.

Results: Fewer neutral responses were noted on the post-survey. One quarter of the nurses in the ED felt the ASQ would be too time consuming to administer, and 33% answered neutral pre-survey. After education was provided, 52% of the nurses felt the assessment would not be too time consuming. Pre and Post survey regarding stigma related to mental health patients remained high at 40%. The number of nurses who felt adequately trained to administer the ASQ 64% post survey, compared to 50% and 14% neutral responses pre-survey.

Discussion: Education on evidence-based suicide screening tool resulted in more ED nurses feeling adequately trained with ASQ education. Continued Mental Health education is needed to assist with decreasing stigma. In the first 3 months of administering the ASQ on all children in the ED ages 10 and over, 390 children were positive in the first 3 months (July 2017 - Oct. 2017). 417 Patients were identified from Oct. 2017 - Feb. 2018.

Category: Evidence Based Practice
Kidney Bucks Program: Incentivizing Compliance in Pediatric Dialysis Patients
Antoinette Thomas, RN, BSN, MBA, CPN; Amanda Marroquin, RD, LD; Amy Sandlin, CCLS; Hanna Boyd, CCLS
Dialysis

Abstract

Background: Background/Purpose. Prior to start of Kidney Bucks program, multidisciplinary team of nurses, dieticians, and child life specialists noted that pediatric dialysis patients struggled with compliance in diet and fluid control and record keeping. Child Life proposed incentives, Kidney Bucks, with education to assist in increasing motivation and thus compliance to address these deficiencies. Purpose of this multidisciplinary quality improvement (QI) project was to increase compliance among pediatric hemodialysis (HD) and peritoneal dialysis (PD) patients and their caregivers, focusing on diet and fluid compliance and record keeping.

Methods: Methods. In February 2015, QI team collaborated with physicians to decide on incentive parameters for phosphorus, potassium, and fluid management. Dialysis dietitian developed and provided ongoing education: one-on-one education, group activities, and teaching skills to appropriately order from a regular diet menu. Data were retrieved through chart audits of labs and clinic visits. Kidney Bucks guidelines are attached.

Results: Results. Of 38 HD patients, 7 were always compliant with phosphorus; 23 patients experienced 60% improvement. For HD patients, 23 were always compliant with potassium, 7 experienced 18% improvement. In both phosphorous and potassium levels, 8 were not compliant. There was very little behavior change for fluid control in HD patients. Of 19 PD patients, 4 were always compliant with bringing PD fluid/urine, home records and procard; 11 showed 58% improvement.

Discussion: Discussion. Current data demonstrated difficulties dialysis patients have with diet compliance and fluid control. Many did not reach phosphorus goal of 3.5-6, but patients improved. This trend was also true with potassium control. To further educate patients, 'Grocery Games' was implemented along with all elements of this QI project. Due to large increase in compliance with record keeping, this focus was removed from project.

Category: Quality
Mechanical Thrombectomy & Catheter Directed Thrombolysis in Children with Severe Cerebral Sinus Venous Thrombosis and Underlying Autoimmune Disease
Jo Tilley DNP, RN, CPNP; Stellena Giddings, LVN, Marcela Torres, MD

Stroke and Thrombosis

Abstract

Background: Despite improved imaging techniques leading to increased recognition, cerebral sinus venous thrombosis in children continues to lack formal treatment guidelines for improved outcomes. Estimated incidence in children is 0.6 per 100,000 children per year (1). Up to 60% of affected children have a focal brain abnormality related to venous congestion resulting in ischemia and/or hemorrhage (2). Poor outcomes, specifically death in 9 to 29% and neurologic deficits affecting primarily cognition and behavior in 50% of affected children, warrants clear evidenced based treatment guidelines (1). 2012 Chest Guidelines support the use of anticoagulation therapy with UFH (unfractionated heparin) or LMWH (low molecular weight heparin) for 3 months with further consideration to extend therapy an additional 3 months based on symptoms and imaging. In severe cases, thrombolysis and mechanical thrombectomy when there has been no improvements with UFH. References: 1. Mongale, P., Chan, A., et al. Antithrombotic Therapy in Neonates and Children: Antithrombotic Therapy and Prevention of Thrombosis, 9th ed: American College of Chest Physicians Evidence-Based Clinical Practice Guidelines. Chest. February 2012 141:2 suppl e737S-e801S 2. Teksam, M., Moharir, M., et al. Frequency and Topographic Distribution of Brain Lesions in Pediatric Cerebral Venous Thrombosis. American Journal of Neuroradiology 2008; 29:1961-1965

Methods: Case review of mechanical thrombectomy and catheter directed thrombolysis for severe CSVT in pediatric patients with underlying autoimmune disease in a single institution from 2014 to 2018.

Results: 3 adolescents with recent diagnosis of autoimmune disorders and extensive cerebral sinus vein thrombosis were all treated with mechanical thrombectomy and catheter directed thrombolysis in addition to systemic anticoagulation therapy with unfractionated heparin (UFH). In Case 1 and 2 the decision to proceed with this treatment modality was based on concerns of suspected antiphospholipid antibody syndrome (APLAS) in setting of severe thrombosis and increased bleeding risks associated with comorbid conditions. In Case 3 worsening neurologic examination and increased cerebral edema requiring external ventricular drain, prompted further intervention with these procedures. In all 3 cases the areas treated with mechanical thrombectomy reaccumulated thrombus within 48 hours of procedure despite continued simultaneous systemic anticoagulation with UFH. However, in all 3 cases vessels with catheter directed thrombolysis using TPA infusion had improvement noted and this was sustained over time in 2 of the 3 cases.

Discussion: Mechanical thrombectomy and catheter directed thrombolysis should remain a therapy option in pediatric patients with severe cerebral sinus vein thrombosis in particular those patients with underlying autoimmune disorders with increased risk of APLAS and worsening neurologic status and worsening neurologic status.

Category: Case Study
Isolated Discontinuous Pulmonary Artery: A case study
Julie Potter MS, Lane Lanier, MD, Vincent Tam, MD

Abstract

**Background:** Isolated pulmonary artery arising from ductal origin (IPADO) is a rare congenital vascular anomaly in which the extrapulmonary portion of the pulmonary artery (PA) is missing with the ductus arteriosus providing continuity with the intrapulmonary vasculature. IPADO may present with concomitant abnormalities such as tetralogy of Fallot or other congenital heart defects. Less commonly, IPADO may be found in isolation. The incidence is 1:200,000. Left untreated, patients may develop pulmonary hypertension, recurrent respiratory infections, and long term significant morbidity including ipsilateral pulmonary hypoplasia and scoliosis. Early and aggressive treatment has shown favorable outcomes in limited, small case series. We report a rare case of an infant with isolated IPADO that underwent successful surgical intervention.

**Methods:** A female infant was born prematurely at 26 weeks gestational age, weighing 1.06 kg, and presented with respiratory failure and hypotension. Worsening respiratory status led to an echocardiogram on day 6, which revealed a patent foramen ovale, bilateral patent ductus arteriosus (PDA), and a isolated right pulmonary artery (RPA) fed by a PDA arising from the innominate artery. Uncomplicated ligation of the left PDA was performed at 17 days of life. Cardiac catheterization, performed at 2 months of age, confirmed the diagnosis and demonstrated supra-systemic right ventricle (RV) pressures without response to oxygen or inhaled nitric oxide (iNO). Subsequently the patient underwent a direct anastomosis between the RPA and pulmonary trunk. Postoperatively, the patient required iNO. iNO was discontinued on postoperative day (PD) #8 after starting Sildenafil. A month after surgery, an echocardiogram demonstrated improved RV pressures at 75% systemic pressure without RPA stenosis. Currently, the 8 month old patient continues to improve with RV pressures 50% systemic. She remains on 0.25L oxygen via nasal cannula and Sildenafil therapy

**Results:**

**Discussion:** IPADO is rarely found in children without associated cardiac defects. Few recent case studies exist and there is no consensus for treatment of DOPA. Management options for this include placement of a Blalock-Taussig shunt (BTS), stenting the PDA or direct surgical repair of the PA. Surgical end to end anastomosis proved successful in this case without residual RPA stenosis, unmanageable pulmonary hypertension or need for further interventions during the hospitalization.

Category: Case Study
Evaluating Cultural Competence in Pediatric Bedside Nurses: Phase 1 Evidence-Based Practice Project Focusing on Culturally Congruent Nursing Care
Jaime Kuhn, BSN, RN, CPN and Jen Bluestein, M.Div.
Nursing/Pastoral Care

Abstract

Background: In 2017, Policy and Procedure Council at Cook Children's Medical Center found Standards of Nursing Practice and Care policy included 13 new nursing competencies for culturally congruent care. First steps were to (1) identify evidence-based cultural competence assessment tool for baseline levels among bedside nurses and (2) develop recommendations to address strengths and deficits in current cultural competence levels.

Methods: Comprehensive literature review using CINAHL and PubMed searches resulted in selection of psychometrically-established self-assessment tool, Inventory of Assessing the Process of Cultural Competence among Healthcare Professionals-Revised (IAPCC-R©). Permission was received to administer IAPCC-R© from developer, Dr. Josepha Campinha-Bacote; IRB approval obtained. IAPCC-R© was administered to 85 nurses, equally spread across critical care, medical/surgical, specialty, and emergency areas. Majority (82%) were baccalaureate-trained, 47% were 20-30 years old, with 46% reporting 0-5 years nursing experience.

Results: From total IAPCC-R© scores, 75% nurses were Culturally Aware; 25% Culturally Competent. Total subscale scores were ranked highest to lowest: Desire, Awareness, Encounters, Skill, and Knowledge. Items with lowest scores were identified: knowledge of ethnic pharmacology (lowest), outside-work involvement with culturally-diverse groups, awareness of cultural assessment tools, definition of cultural competence, and knowledge of biological variations among groups.

Discussion: Phase 2 recommendations to increase the level of cultural competence in nurses include: (1) education blitz, (2) bi-annual cultural competence training using evidence-based self-awareness tool, (3) formation of small groups/journaling and/or unit champions, (4) introduction of cultural assessment tool for admissions, (5) new-hire cultural competency training, and (6) opportunities for community involvement.

Category: Evidence Based Practice
Moral Distress: Effective Interdisciplinary Interventions
Jen Bluestein, M.Div. and Leigh Anne Campbell, MSN, RN, CPN
Pastoral Care/Education

Abstract

Background: Moral distress is commonly reported in healthcare. The purpose of this initiative was to define moral distress, address its impact on emotional and physical wellbeing across disciplines, and expose potential for unsafe patient care. Most moral distress research has focused on critical care and end-of-life healthcare settings. A 26-bed inpatient neurorehabilitation and transitional care unit experienced unrecognized moral distress related to patient complexity. With increasing unit nursing turnover rate (17%) and reported or witnessed outbursts by staff, needs for intervention were vital.

Methods: Moral distress literature points to the efficacy of multi-tiered interventions to reduce effects of moral distress. An evidence-based multi-modal interventional program was implemented over a 6-month period (April-October 2017) by a chaplain and clinical nurse educator. Strategies were selected to increase staff’s knowledge about moral distress, to encourage self-reflection on the experience of moral distress, and to facilitate increased coping skills, including peer support. Interventions included: series of educational videos, online values survey, and facilitated discussion of a case study of a morally distressing patient encounter. Staff were invited to view a PowerPoint presentation reinforcing information shared during case study discussion. Pre- and post-intervention surveys were administered to assess staff knowledge and moral distress experiences and to determine the impact of interventions.

Results: Prior to project, 72% of 61 staff members had never received moral distress training, 85% experienced moral distress during past year, and 49% considered not coming to work or leaving the unit. After program implementation, 96% reported increased knowledge about moral distress and over 80% reported increased coping strategies and abilities to support co-workers during periods of moral distress.

Discussion: This project was an example of interdisciplinary collaboration to tackle the significant issue of moral distress. Continued collaborations would benefit units to increase awareness of the personal, physical and professional impact of moral distress.

Category: Quality
Intranasal ketamine for abortive migraine therapy in pediatric patients: a single-center, retrospective review
Adrian L. Turner, PharmD; Sabrina Shandley, PhD, PMP; Ean Miller, PharmD, BCPPS; Kimberly Tobin, PharmD; Dani Ball, PharmD; M. Scott Perry, MD; Brian Ryals, MD
Pharmacy

Abstract
Background: Dihydroergotamine has been the mainstay of status migrainosus treatment. Ketamine has recently emerged as a promising therapeutic alternative for abortive migraine therapy. Most reports utilizing ketamine for migraine treatment examine adults and the intravenous route. Few reports utilize the intranasal route, and practically none examine intranasal ketamine in pediatric patients. The objective of our study is to further characterize safety and efficacy of IN ketamine for abortive migraine treatment in the pediatric population.

Methods: A retrospective review was performed examining the utilization of intranasal ketamine at 0.1-0.2 mg/kg/dose up to 5 doses in pediatric migraineurs. Pain scores on a scale of 0-10 were recorded at baseline and after each dose. The primary endpoints were efficacy and safety of intranasal ketamine. Response was characterized as pain score reduction to 0-3 and/or reduction of at least 50%.

Results: Forty-one encounters met inclusion criteria. Twenty-nine (70.7%) were responders. Responders had a mean pain score reduction of -6.6. Overall pain reduction from admission to discharge in the entire study population was 57.5%. All side effects were mild and transient.

Discussion: Practitioners should strongly consider integration of intranasal ketamine into abortive migraine therapy algorithms. Our experience has promising outcomes in both pain relief and side effect minimization. Pediatric migraineurs now have multiple, viable treatment options, and intranasal ketamine provides one that is especially desirable.

Category: Research Study
Beyond Magnetic Resonance Angiography in Anomalous Aortic Origin of the Coronary Arteries: Additive Value of Late Gadolinium Enhancement
Steve Muyskens, MD, FSCMR; Andrew Nguyen, OMS-II; Tyler Hamby, PhD
Cardiology

Abstract

Background: Anomalous aortic origin of the coronary arteries (AAOCA) is a common cause of sudden cardiac death (SCD) in young athletes. The prevalence, pathophysiology, and optimal method of risk stratifying AAOCA are unknown. Coronary magnetic resonance angiography (MRA) has been shown to reliably define anatomical features. However, studies evaluating current methods of detecting inducible or chronic ischemia in patients with AAOCA are lacking. We present our institutional experience utilizing late gadolinium enhancement (LGE) as an adjunct to exertional symptoms, exercise stress testing (EST), and single-photon emission computed tomography (SPECT) for risk stratifying high-risk AAOCA.

Methods: A retrospective review was conducted of all patients referred for evaluation of possible AAOCA by cardiac magnetic resonance imaging (CMR) between January 2011 and December 2017. Patients with high-risk coronary anatomy were included; patients with complex congenital heart disease were excluded.

Results: There were 74 patients evaluated for possible AAOCA (median age 14.3 years; 69% male); 40 met high-risk inclusion criteria (34 right, 6 left). SPECT was performed in 33 patients, and EST in 36 patients. Exertional symptoms were present in 11 patients. One patient with aborted SCD had subepicardial LGE, most consistent with myocarditis. No additional patients had baseline ventricular dysfunction or LGE findings on CMR. Risk stratification by exertional symptoms or coronary variant revealed no significant correlation to any markers of ischemia. Furthermore, SPECT was predominantly negative (70%) and 3 of 10 positive results did not correlate with the affected coronary vascular territory.

Discussion: Our study demonstrates the difficulties in utilizing common techniques for risk stratification in patients with AAOCA. While coronary MRA has been shown to reliably assess coronary anatomy, CMR-derived LGE had no additive value in this cohort, and SPECT had a high false positive rate. A larger multicenter study including the utility of stress CMR would be beneficial in this patient population.

Category: Research Study
Effect of an evidenced-based diabetic ketoacidosis pathway in a pediatric emergency department on patient outcomes

Robert Wilson, OMS-II; Rishita Jessu, OMS-II; Tyler Hamby, PhD; Kimberly Tobin, PharmD
Pharmacy

Abstract

**Background:** Our primary objective was to measure time to first insulin dose, before and after usage of a diabetic ketoacidosis (DKA) order set in a pediatric emergency department. We had multiple secondary objectives, including determining the relation of patient outcomes to time to first insulin (e.g., length of stay in the ED, inpatient admissions), and improving time to resolution of all DKA patients.

**Methods:** This project was implemented after an agreement between the Endocrine and Emergency Departments that the establishment of a guideline would be helpful; the ICFISH pathway was created and implemented. ICFISH is an acronym for identify, call endocrinology, fluids, insulin, status, and finally hand-off. The retrospective data collection occurred between October 1, 2014 and March 31, 2016. The prospective data collection was between November 1, 2016 and June 8, 2017. All patients admitted for DKA during this time frame were included, but patients with new-onset diabetes and transfer patients were excluded from analyses. Patient records were entered into REDCap and analyses were performed via SAS (Statistical Analysis Software). Time to resolution, length of stay, whether the patient was discharged from the ED, and whether patient was admitted to PICU were compared based on whether the patient was admitted before or after ICFISH implementation.

**Results:** A total of 198 patients met inclusion criteria, and 127 were excluded, leaving 71 patients. There was a statistically significant decrease in the time to resolution of patients on the floor or PICU from the pre-group to the post-group (p=0.032), as well as length of stay (p=0.004), ED discharge (p=0.028), and PICU admission (p=0.001).

**Discussion:** A standardized order set to treat patients admitted for DKA, via the ICFISH pathway, leads to decreased variability, thus resulting in shorter length of stay in the hospital overall, decreased PICU admissions, and increased ED discharge.

Category: Quality
Bleeding diathesis in a patient with a novel mutation in SERPINF2: A case study of alpha-2 antiplasmin deficiency

K. Elissa Dunlap, OMS-II; Margaret Drummond-Borg, MD; Dorothy M. Adcock, MD; Marcela D. Torres, MD
Hematology

Abstract

Background: Alpha-2 antiplasmin is a serine protease inhibitor that inactivates plasmin and prevents premature breakdown of fibrin clots. Deficiency of this enzyme can lead to spontaneous bleeding and hemophilia-like symptoms, despite normal coagulation and platelet function studies. We present a case of congenital alpha-2 antiplasmin deficiency to emphasize the importance of recognizing a patient with bleeding symptoms despite normal coagulation assays and to report a novel SERPINF2 gene mutation as a cause of this disorder.

Methods: Patient's electronic medical records were reviewed for this case study.

Results: A 7-month-old patient presented with a hemarthrosis of the right knee that was assumed to be septic arthritis, despite negative cultures of the bloody synovial fluid. She experienced significant bruising and spontaneous hematomas, necessitating a hematologic consultation. Complete blood count (CBC), Von Willebrand Factor (VWF) activity and antigen, prothrombin time (PT), partial thromboplastin time (PTT), thrombin time, platelet function analysis, and fibrinogen levels were all normal. At 3 years of age, she had a second suspected hemarthrosis of the right knee, prompting the following tests: plasminogen activator inhibitor-1 levels, euglobulin lysis time, factor XIII activity and platelet aggregation studies. All tests showed normal results. In addition, she had two episodes of hemorrhage after a tooth extraction, both instances required red blood cell and fresh frozen plasma transfusions. Whole exome sequencing revealed a novel homozygous mutation in the SERPINF2 gene. Alpha-2 antiplasmin activity was then measured. Family history was positive for consanguinity. These findings suggest that this mutation has likely been present in several generations of this patient’s family and follows the pattern of an autosomal recessive disorder.

Discussion: This case highlights the difficulties in diagnosing bleeding disorders that involve the fibrinolytic pathway. The previously unreported pathogenic mutation of the SERPINF2 gene may provide valuable insight into the molecular mechanisms of alpha-2 antiplasmin deficiency.

Category: Case Study
Long Term Survival in Pediatric Renal Cell Carcinoma despite multiple relapses: A Case Study
Nidhi D. Patel, OMS-II; Dr. W Paul Bowman, MD; Yvette Dzurik, MD
Oncology

Abstract

**Background:** Although common in adults, Renal Cell Carcinoma (RCC) is extremely rare in pediatric populations, comprising less than 5% of malignant renal tumors. Literature on the topic is scarce, and there currently is no protocol for the treatment of pediatric RCC. This case study aims to highlight pediatric RCC presentation, and provide insight on treatment approaches.

**Methods:** A thorough review of the patient's medical record was performed in order to collect information about the patient's presentation, the treatment approaches, and disease progression.

**Results:** This case focuses on a patient that presented at 6 years of age with painless hematuria, and was diagnosed with type II papillary RCC following pathologic examination of a resected renal mass that had been identified by abdominal ultrasound. Patient underwent a right radical nephroureterectomy, but had several relapses in multiple sites over the ensuing seven years, each of which were addressed on a situational basis with approaches including pneumonectomy, stereotactic radiosurgery, surgical resections, and targeted chemotherapy with Sunitinib.

**Discussion:** In regards to pediatric RCC, a positive nodal status at the time of diagnosis, and recurring metastasis are both indicative of poor prognosis, and since the disease is so rare, there is not a treatment protocol in place. An individual approach on the treatment of this patient has been successful, with the patient being in remission for seven years.

Category: Case Study
Rate of Serious Adverse Events associated with Diazoxide Treatment of Patients with Hyperinsulinism
Paul Thornton, Lisa Truong, Courtney Reynolds, and Jonathan Nedrelow
Endocrinology

Abstract

**Background:** Diazoxide is the first line off label use for hyperinsulinism (HI) and the only Federal Drug Agency (FDA) approved treatment. Its use has increased over the years to include patients with various genetic forms of HI and perinatal stress hyperinsulinism (PShI) and more babies than ever are being exposed to this therapy.

**Methods:** We performed a retrospective analysis of 194 patients with HI in our clinic and looked for those who had suffered serious adverse events (SAE) including pulmonary hypertension (PH) and neutropenia. We compared the rates of serious adverse event in the different types of HI.

**Results:** Out of 194 patients with HI, 165 (84.5%) were treated with diazoxide. There were 15 SAEs in 14 patients (8.5%) including 6 cases of PH and 8 of neutropenia. We found the rate of PH was significantly higher (p=0.023) in those with PShI compared to those with genetic forms of HI.

**Discussion:** The rate of SAEs associated with (not necessarily caused by) diazoxide has been demonstrated. These data show that the frequency of SAE associated with diazoxide use is 8.2% overall but that those with PShI have a much higher rate than those with genetic forms of HI (15.2% vs. 2.4%). Also diazoxide is associated with PH (3.6% of patients treated) and that the association is more likely in patients with PShI (6.1%). This information should help balance the risk benefit of treatment and guide on screening for these complications in the population of treated patients.

Category: Research Study
Isolated Pulmonary Artery Arising From Ductal Origin: A Case Study
Julie Potter MS, Vincent Tam MD, Lane Lanier MD
CICU

Abstract

Background: Isolated pulmonary artery arising from ductal origin (IPADO) is a rare congenital vascular anomaly in which the extrapulmonary portion of the pulmonary artery (PA) is missing with the patent ductus arteriosus (PDA) providing continuity with the intrapulmonary vasculature. IPADO may present with other congenital heart defects. Less commonly, IPADO may be found in isolation. The incidence is 1:200,000. Left untreated, patients may develop pulmonary hypertension, recurrent respiratory infections, and significant morbidity including ipsilateral pulmonary hypoplasia and scoliosis.

Methods:

Results: We present a female neonate delivered at 26 weeks gestational age, weighing 1.06 kg, having respiratory failure and hypotension. Worsening respiratory status prompted an echocardiogram on day 6, revealing a patent foramen ovale, bilateral PDA, and an isolated right pulmonary artery (RPA) fed by a PDA arising from the innominate artery. Uncomplicated ligation of the left PDA was performed at 17 days of life. Cardiac catheterization, performed at 2 months of age, confirmed the diagnosis and demonstrated supra-systemic right ventricle (RV) pressures without response to oxygen or inhaled nitric oxide (iNO). Subsequently the patient underwent a direct anastomosis between the RPA and pulmonary trunk. Postoperatively, the patient required iNO. Sildenafil was started and iNO was weaned to off. A month after surgery, an echocardiogram demonstrated improved RV pressures at 75% systemic pressure without RPA stenosis. Currently, the 8 month old patient continues to improve with RV pressures 50% systemic on supplemental oxygen and Sildenafil therapy.

Discussion: IPADO is rarely found in children without associated cardiac defects. Few recent case studies exist. There is no consensus for treatment of IPADO. Management options for this include placement of a Blalock-Taussig shunt (BTS), stenting the PDA or direct surgical repair of the PA. Surgical end to end anastomosis proved successful in this case without residual RPA stenosis, unmanageable pulmonary hypertension or need for further interventions.

Category: Case Study
Direct Clinical Application with Nurse-Led Research: Impact of 'Boot Camp' Intervention for Caregiver Discharge Readiness
Julie Van Orne, MSN, RN, CPN, Kaylan Branson, MSN, RN, CPN, Mary Cazzell, PhD, RN
Transitional Care Unit

Abstract

Background: Unprepared caregivers for medically-dependent children can result in poor patient outcomes, psychological distress, and financial strain on families and hospitals. A lack of individualized training can lead to inconsistent practice and poor patient outcomes once the patient is transferred home. The purpose of this clinical nurse-led interventional research study was to determine the effectiveness of a 'boot camp' training discharge program on 34 caregivers of 34 medically-dependent children (compared to retrospective data prior to January 2015) as measured by lengths of stay (LOS) on a transitional care unit (TCU) and overall days in the hospital, total discharge teaching days, caregiver stress level, and caregiver satisfaction with training.

Methods: This study utilized a quasi-experimental comparative intervention design and was conducted on the TCU. The 'boot camp' training program included: (1) admission care conference, (2) training contract, (3) structured daily training schedule, and (4) completion with nine training sessions (three hours of content per session). Outcome measurements included: (1) demographic data from 34 caregivers and their children, (2) pre- and post-intervention scores on parental stress from the 36-item Parenting Stress Index-4-Short Form, (3) caregiver satisfaction related to training, and (4) comparisons of average or median LOS of patients on unit and in hospital.

Results: Median unit LOS decreased by 44% from 77 to 43 days; (2) median hospital LOS decreased 44% from 146.5 to 82 days; (3) average discharge training days decreased 62% from 41.5 to 15.7 days; (4) parental Stress scores decreased across all 3 subscales; and (4) caregiver satisfaction with boot camp training showed over 90% were very satisfied. With DRG-related 42-50 day LOS limits, pre-boot camp 77-day LOS, and average daily unit cost of $1974, savings estimated between $53,300 and $69,900 per patient on TCU.

Discussion: The boot camp training model could be customized to other areas where caregiver education is completed.

Category: Research Study
Retrospective analysis of the patients referred to the pediatric nephrology clinic at Cook Children's for evaluation of hypertension
Matthew Pueringer, MS; Jordan Brzezny, OMS-II; Tyler Hamby, PhD; Julie Barrow, RN; Randa Razzouk, MD, MSCI
Nephrology

Abstract
Background: There is increasing evidence that hypertension and pre-hypertension are becoming more prevalent in the pediatric population and may contribute to premature atherosclerosis and the early development of cardiovascular disease and kidney disease. Higher blood pressure in childhood correlates with higher blood pressure in adulthood and the onset of hypertension in young adulthood. Despite the awareness of this increasing prevalence, pediatric hypertension is often unrecognized. The purpose of this study was to quantify the proportion of patients, referred to pediatric nephrology clinics for hypertension, who first had elevated blood pressure (EBP) ≥1 year prior to referral.

Methods: This study was a single center retrospective chart review of patients aged 3-18 years and referred to the outpatient nephrology clinic at Cook Children's for the evaluation of hypertension, pre-hypertension, or EBP between July 1st 2016 and July 1st 2017. We excluded patients referred for diagnoses other than hypertension, patients previously seen by nephrology, and patients lacking appropriate documentation.

Results: Of the 135 charts reviewed, 46 patients met inclusion criteria. Of these, only 6 (13%) were referred within the first year of documented EBP. The median time to referral in years was 3.46 for the population studied. The median number of visits prior to referral with documented EBP (≥95%) was 5, and 9 (20%) patients had ≥10 visits with EBP prior to referral. Cardiology and neurology had high and low rates of referral, respectively.

Discussion: Hypertension, considered a silent killer, is associated with premature atherosclerosis and early-onset vascular disease manifestations. Early diagnosis and management are crucial, and preventative measures to limit further development should be taken. For the present data, both the duration of time and number of visits from first EBP to referral to nephrology are concerning from a prevention standpoint. Further data acquisition and analysis are required but the awareness on this topic is essential.

Category: Research Study
Mid-Term Outcomes of Right Ventricular Outflow Tract Reconstruction Using the Freestyle Xenograft
James A Kuo, MD; Tyler Hamby, PhD; Maham N Munawar, MMS; Eldad Erez, MD; Vincent KH Tam, MD
Cardiology

Abstract
Background: A stentless porcine aortic root (Freestyle) bioprosthesis has been primarily used in our institution for right ventricular outflow tract (RVOT) reconstruction in congenital heart disease. We hypothesize its longevity is similar to other conduits for this indication.

Methods: We performed a retrospective review of all RVOT reconstructions using the Freestyle bioprosthesis at Cook Children’s. Ross procedure, loss of follow-up within a year of surgery, and non-cardiac related deaths were excluded. Survival and reintervention, either by surgery, transcatheter valve implantation, balloon valvuloplasty, or bare metal stent placement were recorded. Factors associated with reintervention were assessed using Cox regression.

Results: Between January 2002 and December 2015, there were 163 patients meeting inclusion criteria. The median age was 12.2 years (range 0.7 to 36.9), median weight was 39.1 kg (range 6.9 to 176), and median body surface area 1.23 m2 (range 0.4 to 2.7). Ninety-three (57%) patients had tetralogy of Fallot. The median follow up was 5.3 years (range 1.04 to 13.7). There were no operative or cardiac related deaths. Thirty-eight patients (23%) required reintervention. The rate of freedom from reintervention was 93.2% (95% CI 86.7-96.6%) at 5 years and 48.4% (95% CI 34.9-60.6%) at 10 years. Age <10 years, weight <39 kg, and body surface area <1.2 m2 at the time of valve placement, as well as valve size ≤25 mm were significantly associated with need for earlier reintervention. Sex, orthotopic versus heterotopic position, diagnosis, or concomitant pulmonary artery augmentation were not associated with earlier reintervention.

Discussion: This is the largest published cohort of primarily pediatric patients with the Freestyle bioprosthesis in the RVOT. It is associated with excellent survival and low mid-term need for reintervention. Its longevity is comparable to published data on homografts and other bioprosthetic valves.

Category: Research Study
Aortic Root Distortion During Balloon Angioplasty of Right Ventricular Outflow Tract Prior to Transcatheter Pulmonary Valve Implantation
James A Kuo, MD
Cardiology

Abstract

Background: Aortic root distortion (AD) can be observed with balloon angioplasty of the right ventricular outflow tract (RVOT), but its long term significance is unknown. This has been a relatively common finding in our institution, though not fully appreciated in our early experience with transcatheter pulmonary valve implantation (TPVI).

Methods: Retrospective review of procedural angiograms prior to TPVI was performed. Degree of AD was classified per grading system and ratio of root diameter change. Valve function and aortic insufficiency (AI) were assessed in follow up echocardiograms post TPVI.

Results: Between June 2012 and October 2017, 47 patients underwent catheterization to attempt TPVI. Five patients had coronary compression which precluded TPVI (one with significant AD as well). Four patients had significant AD and did not receive TPVI (one received bare metal stent alone). Of the remaining 38 successful TPVI, 20 had adequate imaging to assess the aortic root. Four patients had severe AD, 7 had mild AD, and 9 with no AD. The mean ratio of root diameter with AD was 0.70 ± 0.04 for severe AD and 0.85 ± 0.05 for mild AD. Severity of AI did not correlate with degree of AD. Mean follow up after TPVI was 25 ± 21 months. Of the 4 patients with severe AD who received TPVI, 1 has new mild AI with 4 year follow up. Of the 18 patients who received TPVI without adequate arch imaging, 2 patients have new mild AI with 4 and 5 year follow up.

Discussion: AD during RVOT angioplasty is a relatively common finding. Recently, we have not implanted valves in the setting of severe AD. However, in our early experience, this was not well recognized and some patients who were retrospectively identified to have severe AD received TPVI. These patients have done well on follow up.

Category: Research Study
Use of Radiofrequency Energy for Recanalization of Cardiac Baffle Obstruction
James A Kuo, MD
Cardiology

Abstract

Background: Patient is a 39 year old female with d-transposition of the great arteries post atrial switch as an infant. She was noted to have obstruction of the superior vena cava baffle into the heart with decompression by a dilated azygous vein and fistulous connection to pulmonary venous atrium. Novel use of radiofrequency (RF) energy was used to penetrate the obstruction to allow for recannulation with stent implantation.

Methods:

Results: Venous access was obtained via right internal jugular vein and right femoral vein. This allowed for catheters to be placed on either side of the baffle obstruction. This area could not be penetrated with coronary wires or microcatheters. Because of the short segment obstruction, it was felt that RF could be used. Ten watts of energy was applied for 10 seconds through a RF wire. Once this area was penetrated, a coronary guide wire could be passed. The obstructed area was serially balloon dilated and ultimately a stent was placed and dilated to 14 mm. The fistulous connection to the pulmonary venous atrium was occluded using an Amplatzer Ductal Occluder II. Follow up angiography demonstrated unobstructed drainage from the superior vena cava into the heart without shunting into the pulmonary venous atrium.

Discussion: With the expanding adult congenital heart disease program, more adults are receiving care at Cook Children's. Additionally, some have undergone previous surgeries which are rarely performed currently or have unexpected late complications. This can necessitate creative uses of existing technology to accomplish certain procedures. RF is typically used for electrophysiologic ablations. This is our first use of RF to recanalize an obstructed cardiac baffle. Other novel techniques may continue to develop as we care for this complex patient population.

Category: Case Study
Identification of Caregiver Relocation Anxiety from the Pediatric Intensive Care Unit to General Medical/Surgical Units: Utilizing Research to Find Solutions
Danika Meyer, MSN, RN-BC, CPN and Jennifer Horn, MSN, RN, CPN
Organizational and Professional Development

Abstract

Background: Most literature is over 10 years old and focuses on transfers from adult intensive care units to general units. No literature described caregiver relocation anxiety in pediatric intensive care units (PICU). When transfers are viewed as negative, caregivers focus on loss of support from close nurse relationships, change in level of care, lack of predictability in new unit, and perception of type of care on new unit. Purpose was to identify levels of caregiver anxiety across the PICU-to-unit transfer process.

Methods: Prospective repeated-measures design measured caregiver anxiety levels at 3 time points (PICU transfer and 24 hours and 48 hours post-transfer) at large pediatric medical center. Sample included English-speaking primary caregivers, > 18 years, with child in PICU for at least 24 hours. Data collection included: (1) demographic survey and (2) caregiver scores on State-Trait Anxiety Inventory (STAI)8 at 3 time points. Range of STAI scores is 20-80.

Results: 39 caregivers (85% female; mean 36 years; 67% married; 52% high school/some college; PICU length of stay 3 days) completed STAI at all 3 time points (59 total were consented). Descriptive statistics identified moderate 'state' anxiety levels upon PICU transfer (40.8) with decreasing anxiety levels at 24 hours (36.7) and 48 hours (34.2) post-transfer. Repeated measure analysis of variance analyses described statistically significant 'state' anxiety score differences between PICU transfer and 24-hour scores (p=0.025) and between PICU transfer and 48 hours on unit (p=0.001). Post hoc power analysis= 0.98.

Discussion: Study limitations were: (1) Lack of available general unit beds for transfers led to PICU discharges or delayed transfers; (2) Not all eligible caregivers were present at time of PICU transfer; (3) Did not include caregivers transferring to specialty units with existing 'welcome' programs; and (4) Most caregiver participants were female. Identification of caregiver anxiety was essential in developing interventions upon PICU transfer to decrease anxiety and improve satisfaction. Findings support next study to include transfer interventions (unit-specific videos and brochures) resulting in reduction of caregiver anxiety.

Category: Research Study
Caregivers Perspectives on Disclosing a Child's Diagnosis of a Disorder of Sex Development
Cortney Wolfe-Christensen, Kaitlyn Gamwell, Hannah Espeleta, and Larry Mullins
Urology

Abstract

**Background:** Approximately one in 4,000 children will be diagnosed with a Disorder of Sexual Development each year (DSD). Parents are faced with long-term decisions including selecting a sex of rearing, possible genital and reproductive surgery, and hormonal manipulation. The birth of a baby with ambiguous genitalia can be a traumatizing and isolating experience for families. The present qualitative study aimed to understand caregivers' experiences about disclosing their child's diagnosis to others.

**Methods:** Caregivers of children diagnosed with a DSD were recruited across four data collection sites. Thirteen individual, semi-structured interviews were conducted, each lasting 45-60 minutes. Interviews were audio recorded, transcribed, crosschecked for accuracy, and entered into NVivo for analysis. Qualitative data were analyzed utilizing the approach of Patton (2002) in which greater than 80% coder agreement was obtained.

**Results:** The majority of caregivers disclosed details of their child's DSD to immediate family members (77% of participants) and close friends (77%). On rare occasions, people also informed their coworkers (8%), Facebook friends (8%), and individuals involved in child care (8%). Participants reported disclosing information to their family at the child's birth (31%), but telling close friends over time (23%). Concerns about disclosure to others included someone researching the diagnosis, stigmatizing the child, and protecting the child's privacy. When disclosing, they provided vague information (62%). The majority of participants received a supportive response to their disclosure (46%). However, 23% had negative experiences including individuals making harmful assumptions, misunderstanding, or being unsupportive.

**Discussion:** Results demonstrate the complexity of decisions to disclose for parents of children with a DSD. Research supports the vital role of social support in the psychosocial well being of families with a pediatric medical condition. Research should continue to examine how having a child with a DSD may affect social support networks and communication with clinicians, family members, friends, and acquaintances.

Category: Research Study
Health Related Quality of Life in Children Referred for Voiding Problems
Cortney Wolfe-Christensen, Michael Alavi, Kristy Reyes, Blake W Palmer
Pediatric Urology

Abstract
Background: Health related quality of life (HRQOL) is a relatively new concept in pediatrics, but it has shown to be extremely important in terms of clinical care, treatment planning, and compliance with medical and behavioral recommendations. The current project examined whether there were relationships between severity of voiding symptoms and HRQOL in a sample of children referred to Urology for voiding concerns

Methods: Two hundred consecutive patients (74 males, 126 females) between the ages of 5 and 17 (M = 9.2+2.9 years), referred to the Cook Children’s Urology Department for incontinence and/or lower urinary tract symptoms were asked to complete measures of voiding symptom severity (Dysfunctional Voiding Scoring System; DVSS) and HRQOL (Peds QL 4.0 Core Module) as part of their clinical work-up. Both the DVSS and PedsQL are normed and validated measures that have exhibited good reliability. Parents and patients completed the DVSS together, then parents completed the parent-report PedsQL and children over the age of 8 completed a self-report. Since all of the forms have the same number of items and the same subscales, scores were collapsed across age groups for analysis.

Results: Results of correlational analysis indicate that based on parent-report and self-report across the entire sample, more severe voiding symptoms were related to poorer HRQOL in terms of emotional functioning (p<.001; p<.001), social functioning (p<.001; p=.01), school functioning (p<.001; p=.003), total HRQOL (p<.001; p=.01) and the psychosocial health summary score (p<.00; p=.03). Interestingly, HRQOL related to physical functioning and the physical health summary score were unrelated to severity of voiding symptoms (p>.05) on parent-report, but were related on self-report (p=.003).

Discussion: Based on both self-report and proxy-report, children and adolescents with more severe voiding symptoms are at increased risk for having their HRQOL negatively affected across a range of domains.

Category: Quality
Reducing Moderate and Severe Peripheral IV infiltrations and Extravasations

Krystalynn Barre, RN Misty McCutchen, RN
Vascular Access and Wound Care

Abstract

**Background:** With the addition of a new serious harm PIVIE category in 2018, one of the most recent initiatives has been to focus on the prevention of serious harm PIVIES. A trend was discovered through in depth investigation of all serious harm PIVIEs that identified red medications, specifically Calcium Chloride and Calcium Gluconate, as the causality of these events.

**Methods:** Four years of comparison data has been obtained through use of a real-time dashboard that collects data from all Peripheral IV Infiltrations and Extravasations (PIVIE) as well as all PIV Line removals occurring on admitted patients at Cook Children's Medical Center. This data is compared with real-time bedside event reviews and ongoing post-infiltration assessments performed by a dedicated wound care team. The trends assessed through this data are systematically reviewed to determine the effectiveness of implemented initiatives as well as opportunities for improvement surrounding PIVIE reduction.

**Results:** The number of overall PIVIEs has been reduced through the implementation of real-time bedside event review, K-Cards, work instructions, red (vesicant) medication awareness, and patient and family engagement. From 2014 through 2017 there has been a sustained reduction by 7% of total PIVIES. New initiatives have recently been implemented to decrease moderate and severe PIVIES, and most importantly serious harm PIVIE rates to a goal of zero.

**Discussion:** Collaborative efforts have driven the current focus by engaging frontline nursing staff in early identification and recognition and standardized hand-off and documentation of red medication administration and infiltrations. Protocols for early and aggressive treatment of vesicant infiltrations by the Vascular Access Team and Wound Care Team have also played an instrumental role in the ultimate goal of driving down serious harm infiltration rates to zero.

Category: Quality
The Good, The Bad, and The Fever: An Evidence-Based Practice Project for the Treatment of Pediatric Fever within the Urgent Care Center
Shannon Rosiere, BSN, RN, Jennifer Vaughan RN
Urgent Care

Abstract

Background: Fever phobia results in unnecessary visits to urgent care centers (UCCs), contributing to long waits and overcrowding. Surprisingly, fever phobia is not limited to caregivers; multiple studies have reported phobia among healthcare professionals, specifically nurses. Current fever management protocol within a group of pediatric UCCs dictates fever treatment and temperature normalization for all children presenting with fever greater than 38 degrees Celsius. Aftercare instructions given to caregivers of febrile children presenting to UCCs also recommend temperature normalization by alternating antipyretics.

Methods: Iowa Model Revised6 provided guidance from literature review through planning and proposed implementation and evaluation for this nurse-led EBP project. Planning included use of literature on fever management guidelines, symptomatic treatment, and use of antipyretics in the pediatric population which contributed to creation of Fever Management One-Sheet for staff. Consultation with the UCC quality improvement and research council, policy review committee, and education department occurred. Next implementation/evaluation steps in Fall 2017 include: (1) 20-item fever knowledge survey will be given pre- and post- education to staff and (2) Education provided at staff meeting using interactive scenarios and PowerPoint presentation.

Results:

Discussion: Symptomatic fever management protocols in UCCs could be beneficial in increasing fever knowledge and decreasing fever phobia in staff; subsequently decreasing fever phobia among caregivers of febrile children within communities.

Category: Evidence Based Practice
Application of Lean Six Sigma Methodology to Reduce False Positive Venipuncture Blood Cultures in the Emergency Department
Teri Savage, Jonathan Lewis, Megan GraichenKeffer, Heith Gammons, Maggie Higgins, Elizabeth Baker, Dr. Shernette Kydd, Christina John, Traci Baun, Melissia Hampton
Process Improvement

Abstract

Background: The target rate for false positive blood cultures is 3.0% (cited by ASM and CLSI). CCMC had an internal average monthly target rate of 2.0%. The CCMC Emergency Department had the highest contamination rate across the hospital in September 2017, a monthly average of 3.36%. A formal Process Improvement project was initiated to identify sources of variation and to implement process improvements to reduce the false positive blood culture rate.

Methods: The Lean Six Sigma phased approach process improvement methodology was applied. The Define phase identified the problem, quantified a process improvement goal and formulated a project team. In the Measure phase the current process was mapped and measured to establish baseline performance. The root cause of the problem was identified in the Analyze phase and included issue identification and prioritization, process variation studies and process capability analysis. The project team identified and tested a process improvement solution in the Improve phase. The team developed a standard process for collecting blood culture specimens and piloted this process using 28 clinicians (RNs and Paramedics). Hypothesis testing was used to verify the effectiveness and impact of the solution to the process. To complete the project, the Control phase adopted the proven solution to all Emergency Department clinicians via training and skills assessment of the process. The process was monitored for a period of time to ensure the improvements were sustaining.

Results: A group of 28 clinicians piloted a standardized process for collecting blood cultures. The pilot ran from December 2017 through May 2018. The blood culture contamination rate fell from a pilot group baseline of 2.37% to 1.07%, a 55% reduction. A two-sample t-test verified the improvement was statistically significant; p-value of 0.046 at 90% confidence. This exceeded both the target goal of 3.0% and the internal CCMC goal of 2.0%. The standardized process was adopted by 300+ Emergency Department clinicians. The post-pilot period from June 2018 through September 2018 have blood culture contamination rates at an overall average of 1.9% per month. The blood culture contamination rate for September 2018 achieved an all-time low for the Emergency Department of 1.3%. It can be concluded the adoption of the standardized process for collecting blood cultures has significantly reduced the number of false positive blood cultures.

Discussion: False positive cultures may cause stress to the patient as well as unnecessary procedures/medications, extended hospital stays, adverse drug reactions, delayed treatment and risk of harm to the patient. These impacts to the patient may lead to increased costs for laboratory testing, medication adjustments, changes in plan of care, increased nursing care and other associated costs to health boards. This project estimated a conservative cost avoidance of $208,000 annually.

Category: Quality
Only Leaves Should Fall: Evaluating the Effectiveness of the GRAF-PIF Fall Risk Assessment Tool

Alice Stratton, BSN, RN

Abstract

Background: Cook Children's Quality Improvement Council data showed that the currently used fall risk scale, the General Risk Assessment For Pediatric Inpatient Falls (GRAF-PIF), was not sufficiently predictive of falls. Of the patients who fell at the medical center in 2017, 41 were categorized as low risk while 42 were categorized as high risk. The purpose of this evidence-based practice project was to compile a synthesis of evidence-based fall risk factors in pediatric inpatients in order to identify if there is a more valid pediatric fall risk screening tool available for use at Cook Children's.

Methods: A comprehensive literature review and synthesis was conducted and evidence-based pediatric fall risk factors and existing pediatric fall risk assessment tools were identified. The tools were evaluated based upon how many factors they included, whether they had undergone validity and precision trials, and whether they were available for public use.

Results: Three of the seven pediatric fall risk scales reported sensitivity data: GRAF-PIF was 75%, Humpty Dumpty Fall Risk Scale (HDFRS) was 85%, and CHAMPS was 75%. The HDFRS was the most accurate tool in identifying high-risk patients who fell. The HDFRS reported interrater reliability (>70%) with no reported reliability data for GRAF-PIF. Of the seven fall risk screening tools evaluated, HDFRS was found to include the highest percentage of fall risk factors (69%). HDFRS was the only tool to include clear nursing interventions for calculated scores. HDFRS was recommended for practice change for fall risk assessment.

Discussion: After presenting to nurse leaders a task force was created to pilot validation and reliability trials using HDFRS. Replacing GRAF-PIF with HDFRS is expected to lead to improvements in patient safety by more accurately predicting patients at risk for falls and empowering bedside nurses to intervene appropriately before falls occur.

Category: Evidence Based Practice
Changing the Culture of Pain Control in the NICU

Jennifer Guenther

NICU

Abstract

**Background:** Repeated exposure to painful stimulus in the NICU causes the premature brain to develop abnormally. Providing non-pharmacological pain prevention, along with medication guidelines for procedures, will help prevent the long term consequences that arise from uncontrolled pain. Parent perception of their infants care will also improve through involving them in the non pharmacological interventions used on their baby.

**Methods:** A multidisciplinary team introduced background and non-pharmacological methods of pain prevention to all staff during training. Pain medication guidelines were developed by neonatologist and pharmacy reps and introduced in subsequent trainings. The last strategy was the recent implementation of a non pharmacological Comfort Menu to incorporate parents.

**Results:** Improved awareness and consistent pain prevention use is helping to change the culture of pain in our unit. Staff are more comfortable addressing pain and using various prevention methods. Non-pharmacological methods coupled with medication guidelines have helped decreased usage of opioids in our unit. NRC picker scores will be monitored for improved parent scoring on comfort question.

**Discussion:** Continued reinforcement of education for staff and parents will be needed to maintain change of culture related to pain control in the NICU. Further development of new methods will expand our ability to help our patients and families.

Category: Quality
A Quarterly Peer Support Program for Individuals with Bladder Exstrophy and their Families
Emily Margaret Haddad LCSW
Urology

Abstract

Background: The Bladder Exstrophy-Epispidias Complex are rare conditions that affect urinary continence, kidney health and sexual health. Our interdisciplinary team working in pediatric urology hypothesized that peer support, education, and regular programming may improve the overall outcomes for these individuals and their families. We designed a quarterly peer support program and invited every affected adult and child in our database to attend (n=104) along with their siblings, parents, family members and significant others. Disease specific chronic illness support groups can improve risks for isolation, build community, instill hope, provide opportunity for altruism and education, and enhance overall well-being. We found that engagement in this disease-specific support group benefited participants and the ratings of satisfaction were high.

Methods: Every individual with bladder exstrophy (n=104) was invited to participate with their families. The support groups were lead by our social worker and child psychologist. They took place quarterly and participation was free of charge. A focus group was conducted during a session and a survey was submitted to participants after four sessions to rate their experience.

Results: Participants for our bladder exstrophy support group ranged from 2 weeks old to 34 years old for affected individuals. Including family members, on average 29 individuals attended each session. Families traveled as far as 362 miles to attend a session. The overall rating of the support group was 87% with the teen retreat rating to be the highest with a 94%. Families shared in the survey that they felt either less alone or the same after attending the support group. All the families reported that the best part of the support group was meeting other families.

Discussion: Disease specific support groups build community, provide an opportunity for education and altruism, reduce isolation, and enhance overall well being. The bladder exstrophy support group appeared to accomplish those goals for participants.

Category: Evidence Based Practice
Early speech development in Koolen de Vries syndrome limited by oral praxis and hypotonia

Angela T. Morgan, Leenke van Haaften, Karen van Hulst, Carol Edley, Cristina Mei, Tiong Yang Tan, David Amor, Simon E. Fisher, & David A. Koolen

Rehabilitation

Abstract

**Background:** Communication disorder is common in Koolen de Vries syndrome (KdVS), yet its specific symptomatology has not been examined, limiting prognostic counselling and application of targeted therapies.

**Methods:** Twenty-nine participants (12 males, 4 with KANSL1 variants, 25 with 17q21.31 microdeletion), aged 1.0-27.0 years were assessed for oral-motor, speech, language, literacy, and social functioning.

**Results:** Early history included hypotonia and feeding difficulties. Speech and language development was delayed and atypical from onset of first words (2; 5-3; 5 years of age on average). Speech was characterised by apraxia (100%) and dysarthria (93%), with stuttering in some (17%). Speech therapy and multi-modal communication (e.g., sign-language) was critical in preschool. Receptive and expressive language abilities were typically commensurate (79%), both being severely affected relative to peers. Children were sociable with a desire to communicate, although some (36%) had pragmatic impairments in domains, where higher-level language was required.

**Discussion:** A common phenotype was identified, including an overriding 'double hit' of oral hypotonia and apraxia in infancy and preschool, associated with severely delayed speech development. Remarkably however, speech prognosis was positive; apraxia resolved, and although dysarthria persisted, children were intelligible by mid-to-late childhood. In contrast, language and literacy deficits persisted, and pragmatic deficits were apparent. Children with KdVS require early, intensive, speech motor and language therapy, with targeted literacy and social language interventions as developmentally appropriate. Greater understanding of the linguistic phenotype may help unravel the relevance of KANSL1 to child speech and language development.

Category: Research Study
Respiratory Therapists Driving the Pathway: Bronchodilator Frequency Weaning for Inpatient Asthma Patients
Jennifer Horn MSN, RN, CPN; Mary Bina MS, RT-NPS; Jason Reese BSRC, RRT-NPS

Quality

Abstract

Background: The purpose of this clinical practice change was to modify the existing CHAT inpatient pediatric asthma pathway to encourage weaning of bronchodilator frequency based on patient assessment driven by respiratory therapists (RT). Inpatient pediatric asthma patients were being managed inconsistently by the physicians, and bronchodilator frequencies were often weaned only during physician rounds. The overall goals were to decrease length of stay and hospital readmissions.

Methods: In the EBP project, the research team looked at the literature and best practices at peer hospitals to modify the existing pathway. In the modified pathway, RTs wean bronchodilator frequency based on assessment of the patient’s Clinical Respiratory Score (CRS). The evidence-based CRS includes respiratory rate, breath sounds, use of accessory muscles, mental status, oxygen saturation on room air, and color. Based on the CRS, the pathway guides the RT to wean or maintain the frequency of bronchodilator treatments. The pathway was redesigned in the spring of 2016 and piloted on the short stay unit in July 2016.

Results: Three months following the pilot, the length of stay for inpatient asthma patients decreased from an average of 25.8 hours to 24.3 hours, and the 14-day readmission rate reduced from 1.8% to 0.7%. Based on the outcome data, the pathway was implemented on all inpatient asthma patients on hospitalist service in April 2017 with data pending. Interrater reliability of the CRS was monitored throughout the roll-out.

Discussion: This pathway can serve as a guide for other pediatric hospitals to implement bronchodilator frequency weaning pathways based on patient readiness. This RT-driven bronchodilator weaning pathway increased patient safety and weaned patients more efficiently leading to a decrease length of stay and readmissions.

Category: Evidence Based Practice
Association of Quetiapine Therapy with Electrocardiographic Changes in Pediatric Patients Following Surgical Repair of Congenital Heart Defects

Olga L. Rodriguez, PharmD; Johnbosco Umejiego, MBBS, MS, MPH; Lane Lanier, MD
Cardiac Intensive Care Unit

Abstract

Background: It is unknown if quetiapine prolongs QTc interval in patients with congenital heart disease (CHD). Patients with CHD are at high risk for developing arrhythmias due to the underlying cardiac anomaly or surgical procedure. In our center quetiapine is used to augment sedation and treat delirium. We describe quetiapine's effects on QTc interval in patients with CHD.

Methods: Approval was obtained by the institution's Institutional Review Board. It is a pilot observational study of patients diagnosed with CHD that were prescribed quetiapine while in the cardiac intensive care unit (CICU). Electronic medical records were used to collect data from November 1, 2013 to April 30, 2017. Data collected included: age, weight, body surface area, sex, diagnosis, surgical procedure, presence of ventricular arrhythmia, quetiapine treatment, QTc interval, and concomitant QTc prolonging medications.

Results: Thirty-two patients were identified with a mean age of 36 months. The mean ordered dose for quetiapine was 1.26 mg/kg/day. Eighty-eight percent of the patients were also prescribed methadone with a mean dose of 0.45 mg/kg/day. Forty percent of the patients received additional medications with a known risk for Torsades de Pointes. The mean QTc interval before surgery was 429.8 ms (SD: 39.2) compared to the QTc interval during administration of quetiapine 456.9 ms (SD: 41.6). There were no statistically significant changes in the QTc interval in the sample during follow-up. In addition, there were no statistically differences between the dosing of quetiapine and methadone in our sample population.

Discussion: Given the dosing of quetiapine with the concomitant use of methadone and other QTc prolonging medications, our data demonstrated no significant prolongation in the QTc interval. Quetiapine may be used as an adjunctive therapy for delirium or sedation in this high risk population without prolonging the QTc interval. However, a randomized controlled clinical trial is needed to better support this conclusion.

Category: Research Study
'Peaking' Their Interest: An Interactive Game-Based Approach to Catheter Associated Urinary Tract Infection (CAUTI) Prevention Education
Katy Yanez, MSN, RN, CNOR & Abbie Garcia, BSN, RN, CPN
Organizational & Professional Development

Abstract

Background: Quality patient care and safety are strategic goals and initiatives of healthcare organizations today. In an effort to prevent Hospital Acquired Conditions (HACs), specifically associated with Catheter Associated Urinary Tract Infections (CAUTIs), education was created. A Jeopardy-style game 'Flushin' CAUTI in the Potty' was created to actively engage staff to enhance knowledge retention, improve adherence and comfort with CAUTI bundles, prevent HACs, and decrease hospital length of stay and patient costs.

Methods: The target audience was acute care pediatric registered nurses and support staff. The game included three categories: general HAC and CAUTI, insertion bundle, and maintenance bundle. Points for categories were awarded as milliliters of urine. Teams used buzzers to answer questions. For correct answers, urine was emptied from the Foley bag according to the topic's designated number of milliliters. After a final question where teams wagered earned points, the team with the pitcher containing the most urine at the end of the game won. Games are successful strategies for improving learning outcomes, engagement, and retention of knowledge. The use of interactive gaming allows for increased attention and an opportunity to test knowledge in a stress-free environment. Dividing staff into teams provides opportunities to actively collaborate and participate both individually and as a group.

Results: Evaluation strategies comprised pre and post-activity survey data. Pre-education, 44% of staff lacked comfort with both bundles. Post-education, 92% of staff were either comfortable or very comfortable with insertion and maintenance bundles. 93% of staff passed the post-test, with an average score of 90%. A passing score was considered an 85 or better.

Discussion: Implications for practice include nurse satisfaction with creative learning methodologies, prevention of CAUTI, and resultant decreased hospital length of stay and costs. Lessons learned from survey data and comments reflected game enjoyment, staff engagement, and appreciation for gaming as an active learning technique.

Category: Quality
Outbreak Investigation: Bacillus Bacteremia in Patients with Malignancy or Bone Marrow Transplantation.
Marc Mazade, MD, Hospital Epidemiologist.
Infection Prevention and Control

Abstract
Background: Bacillus bacteremia was found in several patients with malignancy or BMT at Cook Children's. There were deaths. An outbreak investigation was requested by oncology. Bacillus cereus infections occur in neonates, the immunocompromised, and patients with catheters. Spores present in rice survive cooking. Infections may cause neutropenic colitis.

Methods: IP logs and medical records were reviewed from 7/1/2013 - 7/11/18. Among patients with malignancy or BMT, case definitions were as follows: confirmed case: more than one positive culture from blood or concurrent recovery of Bacillus from a sterile site; probable case: one positive culture and clinical infection; case of contamination: one positive culture with more than one organism or only one positive culture.

Results: There were 4 confirmed cases, 4 probable cases, and 3 cases of contamination. Seven isolates shared similar susceptibilities. Ages were 10 months to 22 years. Diagnoses were ALL (6), AML (2), neuroblastoma (2), and Diamond Blackfan anemia (1). Four had relapsed. Four infections were present on admission. Seven had ANC < 500. Fever (6), abdominal pain (5), nausea and vomiting were seen. Seizure and mental status changes occurred in two. Two were CLABSIs. One patient arrested. Another with meningitis died. Another died while having CNS relapse. Another had multiorgan system failure and focal embolic brain lesion. One occurred the third quarter of 2013, then two clusters occurred that were separated by 15 months. Three confirmed and one case of contamination occurred from the second through fourth quarter of 2015. One confirmed, three probable, and two cases of contamination occurred from the second quarter of 2017 through third quarter of 2018.

Discussion: As cases were sporadic with half present on admission, a single hospital source outbreak was not suspected. Neutropenia was a risk factor. Invasive infection is possible in the setting of neutropenic colitis. Therefore, adherence with dietary restrictions should be examined.

Category: Quality
Pediatric Midstream Urine Collection Methods in the UCC: Post-Intervention Surveys
Robin Henson, DNP RN CPNP-PC, Audra Fulton, MPAS PA-C, Samad Zia, OMS-II, Tyler Hamby, PhD
Cook Children's Urgent Care

Abstract

Background: BACKGROUND Urinary tract infections (UTIs) are one of the most common bacterial infections in the pediatric population. Diagnosing UTIs requires a urinalysis and often a urine culture. Diagnosing a UTI in the toilet trained child requires consistent staff education and materials for specimen collection to avoid risk of contamination. Contamination of a urine sample can be reduced with a policy which includes consistent instructions to patients and provision of the correct materials. A quality improvement (QI) project conducted in June 2017 at Cook Children's Forth Worth Urgent Care Clinic (FW UCC) surveyed patients and parents regarding midstream urine collection (MSUC), and it showed that the MSUC policy was not consistently followed by the UCC staff. Online education and training were then provided to clinical staff to reinforce this policy. The purpose of this QI project is to conduct the same patient/parent survey to determine if staff education improved adherence to the MSUC policy.

Methods: METHODS The patient/parent survey, which was used in June 2017, was administered to patients June 8-July 3, 2018. The survey included questions concerning the UCC MSCU policy. Patients (ages 4-18) who presented with signs of UTI were given the survey after staff instruction was given and urine was collected. The surveys were conducted on site at the FW UCC.

Results: RESULTS Descriptive statistics and inferential analyses were run to determine improvement after implementing staff education. A total of 20 (2017) and 19 (2018) patients were surveyed. Patients in 2018 were significantly more likely to receive instructions to use 3 wipes (p=0.003), receive 3 or more wipes (p=<0.0001), and actually use 3 wipes (p=0.0004). There was no significance in the likelihood of receiving gender-specific instructions, receiving MSU instructions, following gender-specific instructions, or following MSU instructions compared with 2017.

Discussion: DISCUSSION/IMPLICATIONS This survey should be repeated in 2019 and include assessing verbal and written instructions provided by staff. Continual use of preassembled MSU collection packets and frequent UCC staff educational may result in a greater adherence to the MSUC policy. Comparing survey results with UCC urine contamination rates will help guide future QI projects to help decrease MSU contamination rates.

Category: Quality
IMPROVING OGTT SCREENING FOR CYSTIC FIBROSIS: IS EDUCATION ENOUGH?
Esther Giezendanner, Janet Garbarz, Carrie Stradley, Aditi Prabhakar, Heather Bayers, Rachel Hamik
Pulmonary

Abstract

Background: Annual screening for cystic-fibrosis related diabetes (CFRD) is recommended for all patients with CF starting at age 10 (Moran et al, Diabetes Care, 2010; 2697-2708), typically through a 2-hour oral glucose tolerance test (OGTT). Our pediatric quality improvement (QI) group recognized a need to improve OGTT completion rates based on our 2014 Center Specific Registry Report which showed OGTT screening rates below the national average. Objective: To increase the number of CF patients 10 years and older being screened for CFRD in line with CFF recommendations by encouraging completion of an annual OGTT.

Methods: The pediatric QI group reviewed our current process and barriers to completion and identified lack of patient awareness and low motivation to complete the burdensome test as major challenges. To address the lack of patient awareness, the team updated an educational handout given to patients along with the OGTT test order. Dietitians included discussion of the reason for the test and what it entailed during the annual clinic visit. Additionally, staff made a reminder call if the test was not completed within 4 weeks and addressed any barriers the family cited such as resending lost orders or helping patients locate a convenient lab where the test could be performed. For patients who completed the test, a small prize ($15 gift card or a pair of movie tickets) was provided at the following clinic visit. In addition, staff checked if admitted patients who had been unable to complete an OGTT outpatient met criteria to be scheduled while inpatient. The goals and components of the intervention were presented to staff in all-clinic and nursing staff meetings. The intervention was started in August 2015 across both pediatric and adult programs. It continued through 2016, and is now standardized into clinic practice. Process measures tracked whether the order with handout was given at patient’s annual visit, whether follow-up calls were made if needed, and if prizes were given at follow-up visits. The primary program outcome was an increase in eligible patients completing their OGTTs.

Results: OGTT completion rates among patients improved from a baseline of 25% in 2014, to 35% in 2015, 59% in 2016 and sustained at 55% in 2017. Data was also evaluated by age group category (see chart). Process measures throughout 2016 indicated good compliance with providing the order and accompanying handout (95%), consistency with follow-up calls (78%) and distribution of prizes (64%) was affected by staff turnover.

Discussion: Increasing patient awareness and reminders of the need to complete OGTT coupled with a small prize for completion successfully improved OGTT rates among patients in our pediatric program. When possible, patients also found it convenient to complete the test during a hospital stay. Remaining challenges are the inherent inconvenience of a 2-hour fasting outpatient lab especially given the already high burden of CF care, and maintaining staff engagement in reinforcing the OGTT for patients.

Category: Quality
Abstract

**Background:** Staff and patients both recognized that our annual evaluation visits were chaotic and time-consuming, so our Quality Improvement (QI) group chose to focus on this issue as part of the VIP-F2 program. A time study showed an average downtime (patients in clinic but not with staff) of 54 minutes. Our project targeted decreasing downtime to improve clinic efficiency and patient satisfaction.

**Objective:** To reduce the length of the annual visit in the pediatric CF clinic by decreasing the amount of downtime by 25% within 3 months

**Methods:** The QI group identified that confusion among staff about patient availability resulted in stretches of time when staff could have been with patients but did not recognize the opportunity. In order to address this issue, the clinic implemented red/green tags and a check-off sheet. At the beginning of the annual visit, Medical Assistants (MAs) placed red/green tags at the door of each patient's room. Staff would then turn the tag to red when with the patient and back to green upon exiting so that other staff could recognize the patient was available. Additionally, each staff member was to notify the next person in line to see the patient and check-off sheets were used to determine who still needed to see the patient. Priority access was given to certain staff members in order to ensure smooth clinic flow. Process measures tracked whether staff were using red/green tags and check-off sheets and notifying the next staff member. A week-long time study was repeated monthly to track changes in patient downtime.

**Results:** Significant improvement was noted in downtime, from an average of 54 minutes to 27 minutes (a 48% reduction) after 3 months of implementation of the new process. Total visit time and total wait time also decreased (30% and 49% respectively).

**Discussion:** Total clinic time at annual visits was successfully reduced by implementing staff workflow changes, including taking steps to clarify patient availability and having all staff members share responsibility for minimizing patient downtime.

Category: Quality
Safety and Efficacy of Ledipasvir-Sofosbuvir With or Without Ribavirin for Chronic Hepatitis C in Children Ages 6 - 11 years.
Mary Suzanne Whitworth, M.D. et. al
Infectious Disease

Abstract

Background: Currently there are no interferon-free treatments available for HCV-infected patients younger than 12 years of age. We evaluated the safety and effectiveness of the all-oral regimen ledipasvir-sofosbuvir +/- ribavirin in HCV-infected children aged 6 to <12 years.

Methods: In an open-label study, patients aged 6 to <12 years received ledipasvir 45 mg - sofosbuvir 200 mg as two fixed-dose combination tablets 22.5/100 mg once daily with or without ribavirin, for 12 or 24 weeks, depending on HCV genotype and cirrhosis status. The primary efficacy endpoint was sustained virologic response 12 weeks after therapy (SVR12). Twelve patients underwent intensive pharmacokinetic sampling to confirm the appropriateness of the ledipasvir and sofosbuvir dosages.

Results: 92 patients were enrolled (88 genotype 1; 2 genotype 3; and 2 genotype 4), with a median age of 9 years (range, 6 - 11). Most were perinatally infected (96%) and treatment-naïve (78%). Two were confirmed to have cirrhosis, while the degree of fibrosis was unknown in 55 patients. The overall SVR12 rate was 99% (91/92, 95% CI 94 - 100%). The single patient not reaching SVR relapsed 4 weeks after completing 12 weeks of treatment. The most common adverse events were headache and pyrexia. One patient had 3 serious adverse events: tooth abscess, abdominal pain, and gastroenteritis, which were considered to be not related to study treatment.

Discussion: Ledipasvir-sofosbuvir was well tolerated and highly effective in children 6 to <12 years old with chronic HCV.

Category: Research Study
Acute Flaccid Myelitis Among Hospitalized Children in Texas, 2016
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Pediatric Infectious Diseases

Abstract

Background: Background: This is a multisite study of pediatric patients with acute flaccid myelitis (AFM) in Texas during the year 2016 among 6 children’s hospitals. AFM is a poorly understood disease. Data describing pathogenesis, treatment, and recovery are limited. AFM is a central nervous system/spinal cord disease, characterized by limb paralysis, CSF pleocytosis and MRI findings with gray matter edema of spinal cord in previously healthy children, often with preceding or concurrent viral illness. Outbreaks occurred in the U.S. in 2014 and 2016. Prognosis in 2014 was poor; 80-90% with permanent paralysis. There is no known cause, but it is believed infectious or post-infectious; linked to enterovirus D68 in 2014 cases Anti-inflammatory treatment during 2014 did not appear effective and the CDC later advised against use. There is no recommended medical treatment. AFM is not a nationally notifiable condition and therefore, many cases are not reported Study Objective: Form a collaborative database of Texas cases of AFM that occurred during the 2016 outbreak, creating the largest cohort of regional 2016 case data. Research goals included describing disease presentation, treatments used, response to medical intervention, pathogens identified, long-term outcome data, and any variables associated with prognosis.

Methods: Study Design & Setting: Retrospective chart review of hospitalized patients meeting inclusion criteria among 6 Texas children’s hospitals in 4 major metropolitan areas. (Austin, Dallas/Ft. Worth, Houston, San Antonio). This study was IRB approved for all participating hospitals. Inclusion Criteria: Patients age 0-18 years hospitalized between January 1- December 31, 2016 identified by each participating hospital according to the following Texas Department of Health and Human Services and definition of confirmed AFM: An illness with onset of acute focal limb weakness AND a magnetic resonance image (MRI) showing spinal cord lesion largely restricted to gray matter and spanning one or more spinal segments. Variables: Detailed chart review included the following: demographics, patient medical history, prodromal illness, vaccine history, neurologic exam at nadir of illness, imaging of brain and spine, laboratory data, medical treatments, physical therapy, degree of improvement following treatments, and outpatient visits up to 18 months.

Results: 22 cases of AFM were seen in children in Texas in the midst of a second nationwide epidemic of AFM in the US in 2016. Characteristics described in Table 1. A plausible pathogen was identified in 50% of cases (Table 2). 95% (21) showed some degree of improvement as of the last follow up (94% (17) with and 100% (4) without treatment) (fig 3). 45% had recovery of function (ability to perform activities of daily living); 32% had full recovery of strength and function (fig 1) Among the 8 patients with all extremities involved, 6 had significant residual weakness, ranging from flaccidity in one extremity to complete caregiver dependence. One was lost to follow up after discharge. None of the three patients with Enterovirus D-68 made a full recovery, and all three remain completely or largely dependent on caregivers.

Discussion: Our cases were similar to those described across the U.S. in 2014 Clinical outcomes were markedly better than those seen in 2014 Associated viral infections were varied, but included cases of enterovirus D68. We were unable to find a consistent correlation between a specific treatment and improvement or recovery, therefore it is unclear if the use of anti-inflammatory treatment like IV methylprednisolone and IVIG is beneficial. We did not see complications overall due to anti-inflammatory therapy. Patients with all extremities involved or enterovirus D68 identified appear to have poorer outcomes.

Category: Case Study
Integrating Co-Production in Pre-Visit Planning
Stradley, C; Abramovitz, S; Bayers, H; Corne, C; Garbarz, J; Giezendanner, E; Gipson, M; Hackworth, R; Hamik, R; Jolayemi, O; Lang, S; Lavin, S; Shook, P; Wolff, C; Newman, E
Pulmonary

Abstract

Background: Prior to initiating co-production pre-visit planning (PVP), we did interdisciplinary collaborative planning on a weekly basis. During weekly team meetings, each patient was reviewed with input/discussion from physicians, nurses, dietitians, social workers, respiratory therapists, clinical therapist, physical therapist, child life specialist, research nurses, and pharmacist. Including caregivers in preparing for their clinic visit provides a voice that is integral to comprehensive care for our patients. Our March 2018 90 day goal was to contact caregivers for PVP for 75% of all annual evaluation visits and 50% of three of our providers' patients. Our goal for 2018 is to contact all of our patients/caregivers for PVP with upcoming annual evaluations and 50% of all other patients.

Methods: The quality improvement group brainstormed smart change ideas to include our caregivers in the PVP process. As part of our effort to utilize co-production with our families, we reached out to our Family Advisory Council for assistance. Families were contacted via email or phone call 10-14 days prior to their visit for input. Responses were tracked along with results of a post visit survey.

Results: We quickly exceeded out 90 day goal with 100% of 3 MDs' caregivers contacted and are on track to exceed our 2018 goal with 78% of all patients' caregivers contacted for PVP. Although response to PVP has been poor with only 8% of families responding, comments on surveys and in person from caregivers indicated they were pleased to be given an opening to set priorities for the visit. Post visit surveys revealed 54% of caregivers indicated PVP was very or extremely helpful.

Discussion: PVP provides caregivers an opportunity to ask questions/concerns that may be forgotten at the visit. Challenges included time consuming for staff, incorrect emails and new EMR. Automated messages via MyChart may assist with these challenges.

Category: Quality
Anticoagulation Management during ECMO in Children with Massive Bleeding

Jill Pittman, RN, BSN, Jose Luis Olarte-Motta, MD, Olga Rodriguez, PharmD, Danielle Ransonette, RN, BSN, Tammy Elizondo, RN, BSN, And Roberto Caballero, MD

ECMO

Abstract

**Background:** A protocol for the management of children with massive bleeding while on ECMO was developed. This protocol includes the replacement of clotting factors, RBCs, and platelets. Anti-thrombotic agents are promptly initiated. The ECMO circuit is treated with a low dose heparin infusion which varies depending on the circuit size, infant or pediatric/adult. AT III is infused at 20 units/hr for either circuit. Anti-Xa levels are monitored and is normally < 0.1 units/mL. We maintain the protocol until the massive bleeding resolves.

**Methods:** A retrospective chart review from 2015-2018 included patients on ECMO with massive bleeding (>4 mL/kg/hr) and in which the protocol was implemented.

**Results:** We identified 11 patients with ages 1-17 years old. The infant circuit was used in 5 patients and the pediatric/adult circuit in 6 patients. Four patients had central VA cannulation, 2 patients had VV cannulation and 5 patients had peripheral VA cannulation. The causes of bleeding included major trauma (1), cardiovascular surgery (6), GI bleeding (2), complications from VV cannulation (1), and pulmonary hemorrhage (1). Five patients required surgical intervention to resolve the massive bleeding. During the protocol none of the patients experienced the loss of the ECMO circuit or major circuit clotting. The protocol duration ranged from 7 to 180 hours. The mean bleeding rate decreased from 11 mL/kg/hr to less than 2 mL/kg/hr within the first 12 hours after the protocol initiation. Limitations of the study was to determine the amount of blood products transfused during massive bleeding and the inability to compare to controls.

**Discussion:** Maintaining an ECMO circuit with a low dose heparin infusion and antithrombin III while replacing clotting factors and platelets in patients with massive bleeding is feasible and can be implemented safely. The use of this protocol could potentially decrease the amount of blood products in patients with massive bleeding during ECMO.

Category: Case Study
Development and implementation of a multidisciplinary guideline for pain and sedation in the neonatal intensive care unit
Ellen Bickel, PharmD, Kimberly Tobin, PharmD, David Riley, MD, Artee Gandhi, MD
Neonatal Intensive Care Unit (NICU)

Abstract

**Background:** Literature regarding the use of analgesic and sedative medications in the neonatal population is limited and conflicting. Inadequate pain and sedation control impairs neurodevelopmental outcomes, altering pain thresholds and physiologic responses. The FDA issued a warning in December 2016 regarding use of opioids and benzodiazepines in this population due to concerns of poor neurologic outcomes. A multimodal treatment approach to manage pain is recommended. The aim was to quantify the number of patients requiring methadone pre- and post-implementation of the guidelines and quantify use of all analgesics and sedatives.

**Methods:** Retrospective chart review of NICU patients admitted in 2017. The guidelines were implemented in a staged approach throughout 2017. The number of neonates receiving at least one of the following medications: methadone, other opioids, dexmedetomidine, benzodiazepines, clonidine, gabapentin, intravenous acetaminophen, or intravenous ketorolac during their stay were calculated monthly based on admission date. Patients were excluded if they transferred out of the NICU, received medications for compassionate care only, or were diagnosed with neonatal abstinence syndrome. Data was compiled into pre- and post-implementation groups (pre: 720 patients admitted January through August and post: 379 patients admitted September to December) for analysis.

**Results:** Of the 1,099 patients admitted, 13.9% (153 patients) were treated with one or more of the medications. A statistically significant decrease was seen in the number of patients requiring methadone pre- and post-implementation, \( p = 0.0386 \). A clinically significant decrease in peak daily doses (mg/kg/day) of opioids, \( p = 0.0615 \), as well as a statistically significant decrease in the use of benzodiazepines, \( p < 0.0001 \), and increase in the use of dexmedetomidine, \( p < 0.0001 \).

**Discussion:** The implementation of a multidisciplinary pain and sedation guideline within the NICU resulted in decreased use of methadone, likely secondary to decreased daily doses (mg/kg/day) of opioids. The NICU has also seen decreased use of benzodiazepines and increased utilization of dexmedetomidine for sedation.

Category: Quality
Standard Uptake Value Ratios are not The Best Technique to Differentiate Focal from Diffuse HI.
Endocrinology

Abstract

**Background:** Currently, the use of the 18F-DOPA PET/CT imaging is the most effective technique for identifying and locating focal lesions in patients with congenital Hyperinsulinism (HI). There are two ways to interpret the images; using standardized uptake values (SUV) or looking at the images and visually inspecting for altered areas of uptake (visual method). Data in the literature proposes that an SUV ratio of >1.3 is indicative of focal disease and a ratio < 1.3 suggests diffuse disease. Some surgical centers do not operate if the data suggests diffuse disease. The aim of this study is to determine if using the SUV ratios of >1.3 accurately differentiates focal from diffuse.

**Methods:** In the SUV method, scans are taken at 20, 30, and 40 minutes. Ratios are calculated by taking the max SUV value and dividing it by the second highest value. In contrast the visual method involves looking at the PET scan and determining visually if there is an area higher than all others. If there were discordance between 3 scans, the highest SUV ratio was used. The pathology and SUV ratios were then compared to determine the sensitivity and specificity of the SUV method.

**Results:** To determine the accuracy of the SUV method, we excluded 11 subjects who did not have surgery, 1 subject with an extensive focal lesion (>90% of the pancreas), 2 with Localized Islet Nuclear Enlargement (LINE) pathology, and 3 with pathology consistent with Beckwith-Wiedemann Syndrome. Of the remaining 33 subjects, there were 24 focal and 9 diffuse based on final pathology. Visual inspection of the PET scans allowed 19 out of 22 of the focal lesions to be identified at surgery and cured with minimal resection. Of the 24 focal subjects, 15 had peak SUV ratio of > 1.3. Using > 1.3 as the cutoff point, the sensitivity of the test to detect focal disease is 63% with a specificity of 78% a positive predictive value of 88%.

**Discussion:** Using the SUV value at a cutoff of > 1.3 means that about 37% of our patients would not have been operated on had if we made determination for surgery based on the SUV reading alone. Based on our results, we find visual PET read is superior to SUV max ratio. PET CT should NOT be the decision maker as to whether a patient needs surgery or not.

Category: Research Study
One-year experience with Spinraza in Spinal Muscular Atrophy
Emily Hazen OMS-IV, Marcie Baldwin, MS, CPNP, Angela Pomykal, MSPT, Warren Marks, MD, Stephanie Acord, MD
Neurosciences

Abstract

Background: Spinal Muscular Atrophy (SMA) is an autosomal recessive disease that results in motor neuron degeneration in the spinal cord and subsequent weakness and hypotonia. Spinraza (Nusinersen), an antisense oligonucleotide, is the first and only FDA approved treatment. Data on clinical outcomes and side effects have been limited to a small number of study participants. The purpose of this study was to evaluate the changes in motor milestones, adverse events, and complications in patients who have received Nusinersen.

Methods: IRB approved retrospective chart analysis of all patients (n=28) receiving Spinraza at CCHCS. Laboratory levels were obtained per medication package instructions. Motor outcome was assessed using the Hammersmith and CHOP INTEND motor scoring systems.

Results: Motor milestones improved in 12 patients, remained stable in 13, and slightly decreased in 3. No patient lost functional motor skills. Most patients experienced a subjective increase in daily function. Three of nine patients on chronic ventilator support tolerated significant setting weans, two maintained settings, and only one required ICU care after 2nd dose. Nine patients had prior spinal instrumentation. General anesthetic was used for 64/119 procedures. No clinically significant laboratory abnormalities were noted. Patients below age 7 years had primarily respiratory and swallow dysfunction. Five older patients had procedure related complications. A single patient twice developed post LP headaches requiring brief hospitalization.

Discussion: Respiratory and swallowing difficulties were higher in younger patients and those with Type 1 SMA. Procedure-related complications were seen in older patients and those with previously instrumented spines. This demonstrates the need for exploration of alternative delivery systems such as implanted reservoirs. The lack of laboratory abnormalities suggests the frequency of laboratory testing may be decreased. Improved or stabilized motor control and subjective increased functionality in most patients indicates Spinraza is an effective medication. The maintained or reduced level of ventilatory support in several patients suggests Spinraza improves muscular control of breathing. Overall, Spinraza is safe and effective in SMA Types 1, 2 and 3 even when started in young adulthood.

Category: Research Study
Mithramycin Induces the Efficacy of Etoposide in Preclinical Model for Ewing Sarcoma
Anish Ray, MD; Umesh T. Sankpal, PhD; Lina Albeer; Abigail Hunter, OMS-III; Holly Lout, OMS-III; Kathryn Dunlap, OMS-III; W. Paul Bowman, MD; Riyaz Basha, PhD
Hematology & Oncology

Abstract

**Background:** A fusion protein and transcription factor, EWS-FLI1 is over-expressed in >85% of Ewing Sarcoma (ES) tumors. High throughput screening with ~50,000 compounds identified Mithramycin (MIT) as an inhibitor of EWS-FLI1. A previous clinical trial to test the efficacy of MIT was not successful presumably due to toxicity from the dose required to inhibit EWS-FLI1. We hypothesize that the efficacy and safety of treatment can be enhanced if MIT is used along with standard chemotherapeutic agents. Accordingly, preclinical experiments were conducted to test combination treatment with Etoposide (ETO).

**Methods:** ES cells, CHLA10 and TC205 were treated with optimized doses of MIT or ETO or MIT+ ETO. The effect of these treatments on cell growth inhibition was studied using a luminescent (CellTiter-Glo®) cell viability assay. The effect of these treatments on programmed cell death was measured by evaluating the apoptotic cell population using Flow cytometry with Annexin-V Apoptosis Detection Kit and the expression of Cleaved-Poly (ADP-ribose) polymerase (c-PARP), by Western blot analysis. Nude mice were injected with TC205 cells, treated with MIT or ETO or MIT+ETO and the tumor growth inhibition was assessed.

**Results:** As expected, the combination treatment of MIT+ETO was significantly (p<0.01) more effective than individual agents. The cell growth inhibition in combination treatment was accompanied by an increase in apoptotic cells and c-PARP expression in both ES cell lines. The combination treatment significantly (p<0.01) decreased the tumor weight and volume when compared to control or any single agent.

**Discussion:** The combination of MIT+ETO is effective when compared to individual treatments in the TC205 and CHLA10 cell lines and inhibited tumor growth in a mouse model. These results demonstrate that MIT in combination with the chemotherapeutic agent ETO increases therapeutic efficacy in preclinical model for ES and demonstrates strong potential for translational application.

Category: Research Study
Re-Solving Resuscitation Response in Patients with Do No Attempt Resuscitation (DNAR) Orders
Lauren Walter, BSN, RN, CPN & Rebecca Ryan, BSN, RN, CPN
4 Main - Surgery & Trauma Services

Abstract

Background: PICO question was: Does creating a standardized method for identifying patients with Do Not Attempt Resuscitation (DNAR) orders increase staff awareness of resuscitation status? Nurse residents at a pediatric medical center developed and implemented an evidence-based practice (EBP) project to address an event where a patient's DNAR status was unclear; resuscitation was inappropriately attempted. Many hospitals have found their current policies regarding DNAR status identification allowed for miscommunication and wrongful resuscitation (‘Color-coded wristband...,’ 2006; 'Initiative sparked..,' 2007; Marcus, 2015; Sehgal & Wachter, 2007; Schiebel et al., 2012; Traynor, 2009; Watson, 2009). In a survey of Colorado hospitals, 70% of respondents identified situations where DNR orders were confusing and led to patient care problems ('Color-coded wristband...,' 2006).

Methods: Current policy at a pediatric medical center required DNAR stickers placed on paper charts and DNAR status identified under Critical Information in electronic health record. Because charts are not kept at bedside, there was no standard bedside identification of DNAR status. Iowa Model Revised (University of Iowa Hospitals and Clinics, 2015) directed this project: (1) literature review and synthesis; (2) communicating with nurse educators, code team, palliative care team, and pastoral care; (3) five-item pre-survey on effectiveness of current method of DNAR status identification; (4) selection of green code sheet (list of emergency medication doses) as bedside identifier to contrast with white code sheet at bedside; (5) pilot of use of green code sheets on three pediatric subspecialty units; (6) five-item post-pilot survey; and (7) revision of current policy for DNAR status identification.

Results: Pre-survey results revealed almost 50% of nurses experienced situations where DNR status of patient was unclear; in post-pilot survey, 92% of nurses reported they felt the resuscitation status was clear. Close to 50% of nurses reported that the pre-pilot practice and policy were ineffective in identifying DNR status versus post-pilot, 92% nurses reported green code sheets were effective.

Discussion: House-wide implementation recommendations were proposed to shared governance councils such as Nurse Practice Council, Professional Development Council, Nurse Executive Council, and Nurse Manager Council. An education module was created to inform all employees of the updated policy, which was rolled out house-wide in March 2018. This practice change will lead to improvement in patient safety and clearer DNAR status identification for hospital staff.

Category: Evidence Based Practice
Acute Torsion of Extralobar Sequestration: Case Report
Matthew Mitts, OMS-II; Marty Knott, DO, PhD
Pediatric Surgery

Abstract

**Background:** Pulmonary sequestration is a congenital formation that involves the growth of non-functional lung tissue that is not perfused by the lung arterial blood supply. There are two types of pulmonary sequestrations, intralobar and extralobar. Intralobar sequestrations are more common and usually present in adults as recurrent pneumonia. Extralobar sequestrations are less common, asymptomatic, and usually are found incidentally on prenatal ultrasound. Presentations of symptomatic extralobar sequestrations are very rare and are most commonly diagnosed postoperatively.

**Methods:** Electronic medical records were reviewed for a 6-year-old patient with an extralobar sequestration.

**Results:** We describe a case of a 6-year-old boy who presented with a three-day history of acute abdominal pain, tachypnea, and a right pleural effusion. A tube thoracostomy was performed to drain his effusion in attempts to improve his symptoms. Computed tomography revealed a right posterior mediastinal mass that was concerning for malignancy. The decision was made to proceed with surgical excision versus biopsy. The patient was found to have a necrotic extralobar sequestration due to torsion. Thoracoscopic resection was performed and the patient was discharged without complication.

**Discussion:** There are only 10 previously reported pediatric cases of an extralobar sequestration with torsion and only 1 reported on the right side. Extralobar sequestrations appear on imaging as posterior mediastinal masses which elicit a broad range of differential diagnoses in children and are often neoplastic. A review of the pediatric literature demonstrates a common radiographic, pathologic, operative and clinical vignette to maintain a high suspicion of extralobar sequestration for the differential of a paraspinal mass.

Category: Case Study
A Novel Mutation of APOB in Two Siblings with Hypercholesterolemia
Abigail Sprunger, OMS-II; Luke Hamilton, MS; Tyler Hamby, PhD; Don P. Wilson, MD
Pediatric Endocrinology and Diabetes

Abstract

Background: Familial hypercholesterolemia (FH) is a common genetic disorder cause of premature atherosclerosis due to chronically elevated low-density lipoprotein cholesterol (LDL-C) levels from birth. Individuals with FH experience an increased risk of premature cardiovascular disease (CVD), and lack of early identification and treatment increases the risk of CVD-related coronary events later in life. We report two siblings with FH caused by a novel mutation in APOB.

Methods: Electronic medical records were reviewed for two patients with FH.

Results: Two biologically related siblings (male age 9, female age 11) were found to have LDL-C levels >95th centile for respective age and gender. Neither sibling had preexisting medical conditions nor a history of chronic medications. Both siblings were found to have the same missense variant in the APOB gene, a novel mutation causing hypercholesterolemia. Because of parental concerns regarding use of statins, both were treated with a cholesterol absorption inhibitor.

Discussion: Despite the benefits of early identification of those at moderate-to-severe risk, several knowledge gaps impede successful cholesterol screening of children: misunderstanding goals of screening, the best screening method, and ideal age for screening and for intervention. Current guidelines recommend universal cholesterol screening and selective screening starting at 10 and 2 years of age, respectively. Although not routinely preformed, identification of a genetic mutation helps to 1) confirm the diagnosis of FH; and 2) serves as an additional risk factor for CVD, aids risk stratification and clinical-decision making, and helps determine the timing and intensity of treatment that would provide the best long-term health benefits. In addition to lipid-lowering medications, treatment should include global reduction of all CVD risk factors through health education, and adoption of life-long, heart-healthy living with a goal to reduce LDL-C levels to <100mg/dL or at least 50% or more.

Category: Case Study
Delayed Diagnosis of Langerhans Cell Histiocytosis: A Case Series

Anna C. Cooke, OMS-II; Mayme Richie-Gillespie, MD; W. Paul Bowman, MD
Hematology and Oncology

Abstract

Background: Langerhans Cell Histiocytosis (LCH) is a clonal proliferative disease of the dendritic-derived, antigen-presenting Langerhans cells. Formerly termed Histiocytosis X, LCH encompasses a wide array of symptoms that range from solitary bone lesions to multisystem involvement. Definitive diagnosis of the disease is made histologically with positive staining of CD1a or Langerin/CD207, which are proteins specific to the aberrant cells. Because of the variable presentation of LCH, the path to diagnosis can be delayed as it tends to have an insidious onset and is potentially confused with other disease processes.

Methods: Electronic medical records were reviewed for three pre-teen patients with LCH who were initially diagnosed with chronic osteomyelitis.

Results: Three patients (2 male, 1 female) initially presented to their primary care physicians with long-term pain and decreased activity in the absence of other systemic symptoms or trauma. In each case, initial diagnoses of chronic osteomyelitis were given. Following surgery, pathology reports did not indicate the presence of LCH, and all cultures were negative. When CD1a and Langerin staining were applied in retrospect, staining for all three patients was positive in small clusters of cells. Final diagnoses of LCH, fibrosing phase, were eventually given and the prolonged clinical history accounted for the amount of fibrosis present in the tissue samples.

Discussion: Because of the variable presentation of LCH, diagnosis can be delayed as symptoms may be attributed to other diseases in the differential. These cases are important because they bring attention to a potential presentation of LCH as gradually occurring bone pain caused by solitary bone lesions. In addition, correct and early identification of LCH provides a quality of life improvement for patients and their families as early detection prevents the hassle, expense, and danger of unnecessary treatment.

Category: Case Study
Delayed Diagnosis of Ewing Sarcoma: A Case Series
Blake St. Louis, OMS-II; Mayme Richie-Gillespie, MD; W. Paul Bowman, MD; Tyler Hamby, PhD
Hematology & Oncology

Abstract

**Background:** Ewing Sarcoma is the second most common bone cancer in pediatrics, affecting mostly adolescents and young adults between the ages of 15 to 24. Ewing Sarcoma presents with a variety of non-specific symptoms, including pain and possible mass, and the average delay in diagnosis is 6 months. Physicians account for 63.6% of the delays in Ewing Sarcoma. Adolescent (15-19 years old) patients have a significantly longer patient-related delay, contributing to an overall longer delay in diagnosis. Patients diagnosed without metastasis have a 70% survival rate, while patients with metastasis present have a 20% survival rate.

**Methods:** Electronic medical records were reviewed at Cook Children’s Medical Center for three patients.

**Results:** Three cases of adolescent Ewing Sarcoma diagnosed between 2010 and 2018 were evaluated. One patient was diagnosed with sciatica followed by a herniated disc after a motor vehicle accident and had a six-month delay. He had not responded to treatment and was referred to a pain management specialist. The two other patients had delays of about 12 months and were being treated during this time by either a chiropractor or physical therapist. Both patients thought the pain was musculoskeletal pain and delayed going to a physician. Two patients are currently in remission and one patient is currently undergoing treatment for Ewing Sarcoma of the pelvis.

**Discussion:** Ewing Sarcoma presents with nonspecific symptoms and can have lengthy delays in diagnosis. Timely diagnosis is important because longer delays can result in metastasis and poor prognosis. Adolescents have significant delays in diagnosis almost triple that of younger pediatric patients. As demonstrated by these three cases, pain onset, proper imaging guidelines, age-specific statistics, avoiding specialty bias, and response to treatment are important components to developing a well-formed differential diagnosis. We hope to improve awareness and early diagnosis Ewing Sarcoma among all physicians.

Category: Case Study
Efficacy of Sympathetic Nerve Blocks in Pediatric Outpatients with Central Sensitization of Pain
Sterling Blackham, OMS-II; Cole Romney, OMS-II; Artee Gandhi, MD; Meredith Brooks, MD, MPH
Pediatric Anesthesiology

Abstract

**Background:** Central sensitization (CS) is distinguished by an increased sensitivity to painful stimuli, due to a lower pain threshold or an altered response to pain. There is currently no standardized treatment for CS. Therapy is normally attempted through medications, lifestyle modifications, cognitive-behavioral therapy, physical therapy, interventional pain procedures, and/or surgery, but evidence supporting specific treatments is lacking. Interventional nerve blocks are relatively common for treating chronic pain in adults but not children. To our knowledge, this is the largest report on sympathetic nerve blocks for treating chronic pain in children.

**Methods:** This study was a retrospective chart review of pediatric patients (aged 0-21 years) treated between January 2009 and March 2018 for CS at Cook Children's Medical Center. Demographics, diagnoses, pain scores (FACES, VAS, NAS, FLACC), complications, and changes in functionality were reviewed for each patient. Though each patient had 1-5 procedures, study analysis was limited to the first procedure.

**Results:** There were 69 distinct patients (56 females, 13 males; 63 Caucasian; median age 14.68 years, range 3.54-19.27 years). Common diagnoses included complex regional pain syndrome (45 patients) and abdominal pain (15 patients). Most patients demonstrated improvements in leg (77%), hand (76%), and overall functionality (71%). Pain scores improved post-intervention for 74% of patients, and on average, patients reported a statistically significant improvement in the pain score post-intervention, *P*<.0001. Post-intervention complications included weakness (n=1); numbness over abdominal area (n=1); nausea (n=1); and hoarseness (n=3).

**Discussion:** CS can manifest in various diagnoses and can cause chronic pain in pediatric patients. Results suggest that sympathetic nerve blocks effectively and safely treat CS in children. Most patients had no complications and showed improvement in pain and functionality post-intervention. Prospective studies, with standardized pain scales and functionality assessments, should be attempted to further support the effectiveness of sympathetic nerve blocks in pediatric patients with CS.

Category: Research Study
Skeletal dysplasia and growth failure in congenital hypothyroidism due to a novel form of generalized thyroid hormone resistance
Matthew Mitts, Preetam Gongdidi, Luke Hamilton, Don Wilson, Joel Steelman
Endocrinology

Abstract

**Background:** Thyroid hormone exerts systemic responses that encompass a wide array of vital functions, including energy homeostasis, skeletal growth, neural development, cardiac function, and gastrointestinal function. These actions are mediated by binding of thyroid hormone to its receptors, allowing tissue specific functions. There are two thyroid hormone receptors, TR-Alpha and TR-Beta that are each encoded by genes (THRA & THRB) on chromosomes 17 and 3, respectively. Until recently, only mutations in THRB were known and have a known incidence of 1 in ~40,000 with an autosomal dominant inheritance pattern. Mutations in THRA have recently been discovered and are exceedingly rare, with 14 cases documented in the past five years.

**Methods:** We present a case of a young girl with skeletal dysplasia due to a novel mutation affecting thyroid hormone function.

**Results:** She was born at term appropriate for gestational age. Prenatal ultrasound was suspect for short long bones. Genetics consultation after birth confirmed ultrasound concern with finding of rhizomelia. She had post-natal growth failure noted with delayed closure of fontanelle. Myoedematous appearance noted. Newborn screening tests were normal; however, subsequent work-up suspect for central hypothyroidism. Levothyroxine started at 11 months of age. Patient seen in CCMC endocrinology in 2017. She had previously been under the care of different endocrinologists. Continued slow linear growth (SD -4.2) noted even in spite of addition of levothyroxine with extreme developmental delay. Whole exome sequencing completed in 2013 showed THRA mutation. Addition of liothyronine in 2014 based upon recommendations of expert and continued abnormally slow growth.

**Discussion:** Most conspicuous of these defects are the skeletal deformities and growth retardations found in patients with a THRA mutations. We describe a case of a 6 year old female who has a unique thyroid receptor alpha mutation with severe skeletal deformities, characteristic of a THRA mutation. Associated features often include a multitude of other hypothyroid conditions, making the diagnosis of THRA mutations difficult.

Category: Case Study
Branchial Cleft Cyst: A Case Study in a Thirteen Year Old Girl
Nasim Shakibai, OMS-II; Michelle Marcincuk, MD; W. Paul Bowman, MD
ENT

Abstract

Background: Second branchial cleft cysts are the most common branchial cleft lesions, found near the anterior upper one-third aspect of the sternocleidomastoid muscle. Branchial sinuses or fistulae are often found after birth, but internal sinus tracts and branchial cleft cysts may be found later in life. Although branchial clefts are resectable, the surgeries become more complicated following an infection. We report a case of a patient with a second branchial cleft cyst with a previous infection, leading to a difficult resection.

Methods: Electronic medical records were reviewed from a patient with a second branchial cleft cyst.

Results: A 13-year-old female patient presented to her pediatrician with tender mass on the left side of her neck for two days. She denied fever, fatigue, weight loss, or cat scratches. Her boyfriend had Infectious Mononucleosis a few weeks prior. Her labs revealed a normal Epstein-Barr Virus panel and a negative mononucleosis spot test, rapid strep A test, and Bartonella Henselae Antibodies. Her erythrocyte sedimentation rate and C-reactive protein were elevated. She completed a course of clindamycin and azithromycin, but the mass remained. A chest x-ray revealed clear lungs. Her MRI with contrast showed a mass which her otolaryngologist identified as a branchial cleft cyst. The resection was complicated due to her previous infection; the cyst was adherent to the great vessels, the accessory nerve and the sternocleidomastoid. The patient recovered uneventfully.

Discussion: Branchial cleft cysts account for approximately 20-30% of all pediatric neck masses. If a patient presents with a neck mass, health care providers should consider branchial cleft anomalies in the differential diagnosis. Knowledge about branchial cleft cysts and their presentation will help health care providers ensure these patients receive appropriate management more rapidly, thus avoiding some of the challenges and risks of surgical excision once infection has occurred.

Category: Case Study
Diffuse Cutaneous Mastocytosis and its potential comorbidities in pediatric patients - a case study
Kolton Smith, OMS-II; W. Paul Bowman, MD
Hematology and Oncology

Abstract

**Background:** Mastocytosis is the pathologic proliferation and accumulation of mast cells in various tissues of the body. There are different forms of mastocytosis that can present in pediatric patients including systemic (SM), cutaneous (CM) and diffuse cutaneous mastocytosis (DCM). Both the CM and DCM forms have the potential to progress into SM as the patient reaches adulthood. Mastocytosis has been shown to be comorbid with joint pathologies including Ehlers-Danlos syndrome and inflammatory gastrointestinal conditions such as eosinophilic esophagitis. The greatest risk among patients with mastocytosis is anaphylaxis.

**Methods:** Electronic medical records for this patient were reviewed for this case study.

**Results:** A 13-week-old male presented to his primary care physician with erythematous spots on his torso and arms, and was diagnosed with eczema. The spots grew and transformed morphologically over the next month and a referral to dermatology was made. Upon biopsy of the original lesion (on the torso), the diagnosis of DCM was made. Over the coming months, symptoms progressed and comorbidities-including joint hypermobility (diagnosed with Ehlers-Danlos syndrome), dysphagia and diarrhea-arose. The patient broke his distal radius while crawling, due to his mast cell disorder and severe vitamin D deficiency.

**Discussion:** In most children with DCM, symptoms will partially or fully resolve by adolescence. But for some patients, the disease can progress to SM. Numerous comorbidities can occur, as did in this case. Current treatment strategies are wide ranging, from topical glucocorticoids to specialized UV radiating therapy. The specific approaches to this disease are still being understood, with recent investigations into immunological treatment modalities. The individuality of each case is crucial for health care professionals to recognize.

Category: Case Study
SIMV/VG a new mode of ventilation introduced in CCMC NICU. Was it beneficial or not?

Dr. David Riley M.D/Dianne Jones RRT/Julie Kirkwood RRT/Respiratory Educator
Respiratory NICU

Abstract

**Background:** 1. SIMV/VG was a new mode of ventilation that was available to be used in our NICU. 2. The goal was to decrease our patients days on the ventilator and therefore decreasing length of stay in CCMC NICU. 3. In decreasing the number of days our patients spent on the ventilator by using SIMV/VG we anticipated a shorter NICU stay. Long term the babies would have better outcomes and experience less lung damage from days spent on a ventilator.

**Methods:** 1. SBAR was used to communicate our ventilator strategy for using SIMV/VG. 2. Q&A in service was setup to share information with all staff that would be involved in caring for our patients 3. A algorithm was developed as an useful tool that would ultimately help staff with trouble shooting this new mode of ventilation. The NICU Respiratory Terapist would be vital in the success of SIMV/VG.

**Results:** 1. Improved staff communication with all team members. 2. After an initial learning curve most of our NICU patients were immediately placed on SIMV/VG on admission. 3. Final data will be submitted ASAP to determine the success of SIMV/VG in CCMC NICU.

**Discussion:** The ultimate goal of using SIMV/VG here at CCMC was to decrease lung overdistension of the lungs. This in itself can lead to Bronch-pulmonary dysphasia in our patients which can cause long term lung problems for the NICU patients who have been on ventilators.

Category: Research Study