EVALUATION OF AN INTERACTIVE WEB-BASED EDUCATIONAL MODULE FOR HEALTH CARE PROVIDERS TREATING PAIN IN PATIENTS WITH SICKLE CELL DISEASE

by

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Acute pain from vaso-occlusion that cannot be reduced at home is the most common reason for Emergency Department (ED) visits and hospital admission for patients with Sickle Cell Disease (SCD). Administration of opioids is often necessary to alleviate pain symptoms. Despite health-care provider's knowledge of pain episodes that occur in individuals with SCD, poor pain management persists in the health care setting. This may be due to preconceptions and misinformation regarding administration of opioids and general lack of SCD knowledge. The availability of personal computers and access to the World Wide Web in the home and office settings allows for web-based learning as an efficient and convienient method to reach many health-care providers. Clinicians can access concisely presented information that covers the most essential information, and that provides instantaneous access to relevant literature. Consequently, we developed a web-based, interactive educational module that describes a typical presentation and course of a pediatric patient with SCD presenting to the ED with pain. The purpose of this project is to assess whether this type of educational intervention is useful, accessible and effective for enhancing health care provider's knowledge. To measure degree of knowledge gained, we compared answer selections from a 5-question pre-test and post-test. An evaluation was administered containing two demographic questions and nine opinion questions. Opinion questions asked subjects to what degree they felt a particular objective was achieved.

Answers choices were based on a Likert-type scale ranging from “very low” to “very high”. A variety of health care providers have participated in this project, ranging from social workers to hematology attending physicians. Subjects reported feeling a web-based application was useful and that the overall quality of our module was very good. Under-treatment of pain in all patient populations is a public health problem. In the SCD patient population pain episodes recur over a lifetime and significantly impact all areas of life. Web-based educational modules similar to the one we have created can augment knowledge and refine health care providers’ practice of pain management in SCD to improve efficacy, thereby increasing quality of life for these patients.

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PREFACE

It is amazing to me, when I think back to all the classes, clinics, lectures, and projects, how much I have learned and grown during my short time here. I feel so glad and so lucky to have had the opportunity to be a part of such a supportive and exciting program that has provided me with experiences I know would not have had anywhere else. I am grateful to all of the faculty and staff within the Department of Human Genetics for their kindness and encouragement to myself and my classmates over the last two years. I would like to extend a special thank you to Robin Grubs and Betsy Gettig. Their continued contribution to the genetic counseling profession and support to their students is an inspiration.

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1.0 INTRODUCTION

The clinical hallmark of Sickle cell disease (SCD) is acute pain that occurs from vascular occlusion. (Ballas 2005) Pain episodes recur unexpectedly and without warning over a lifetime, often requiring intervention by the Emergency Department (ED) with administration of opioids for alleviation of pain sensation. Despite the development of management guidelines by the Joint Commission on Accreditation of Healthcare Organizations (JCAHO) and the American Pain Society (APS), it is well established that effective pain management continues to be a problem for all patient populations. (Rupp and Delaney 2004; Todd KH 2007) This is especially troubling for patients with SCD. Unlike post-surgical or cancer-related pain, individuals with SCD experience acute pain episodes throughout their lifetime. This leads to developing tolerance to typical doses of opioids due to chronic exposure. Although clinicians are aware of pain episodes that occur in SCD patients, poor pain management practices persist. This often results from a complex cycle of preconceptions and misinformation regarding management of pain with opioids, paired with health care providers' limited knowledge of SCD and failure of institutional guidelines for effective pain management in SCD to be understood by health care providers. (Yadgood, Miller et al. 2000) Clinicians' misinformation is paired with patient anxiety that stems not only from pain sensation but also from anticipation of not being believed by health

care providers. This can perpetuate a cycle of inefficient pain management as tension increases between provider and patient.

The Comprehensive Hemoglobinopathy Program at Children's Hospital of Pittsburgh has provided multidisciplinary care to children with SCD since 1978 and provides care for over 200 children each year. The program is made up of a team of Hematology experts including physicians, nurse practitioners, social workers, and behavioral health specialists. The primary goal of this program is to identify children with SCD early in life and effectively manage their condition. One step in achieving that goal was to create an algorithm that describes a step-wise management protocol to help streamline and focus pain management for patients with SCD presenting to the ED with a pain episode from vaso-occlusion. This algorithm has been implemented into Children's Hospital of Pittsburgh's Guidelines for Clinical Effectiveness (Appendix D).

There is a need to augment health care providers' knowledge regarding effective pain management from vaso-occlusive crisis. Additionally, increased recognition of the signs and symptoms of life-threatening circumstances that can occur secondary to pain episodes will allow clinicians to better understand how to efficiently tackle pain stemming from vaso-occlusive crisis as well as develop effective strategies for treating their patients with SCD. In an effort to promote the algorithm and to provide an opportunity for health care providers to enhance their understanding of Sickle cell disease, we have created an interactive, web-based educational module that follows a patient and his mother through their visit to the ED due to pain from vaso-occlusion. Using the algorithm as a guide, users must answer questions regarding effective management of the patient's pain episode.

The purpose of study was to assess whether an interactive, web-based, educational module would enhance health care providers ability to manage acute pain from vasooclusion in patients with SCD. Our intent was to create an educational module that was concise, accessible, and appealing to the user. By addressing the four aims listed below, we measured both knowledge gained and assessed subjects' opinions of this type of media to improve their knowledge, attitudes and efficacy in caring for SCD patients who present to the ED with acute pain from vasooclusive crisis.

Aim 1: Assess whether health care providers have increased knowledge of management of acute pain from vaso-occlusion in patients with SCD.

Aim 2: Determine whether health care providers will alter they way they manage patients with SCD presenting to the ED or to their clinical practice with pain following the use of this educational module.

Aim 3: Determine health care providers overall attitudes about web-based educational modules.

Aim 4: Assess whether this type of educational tool helped health care providers to learn and understand the established CHP algorithm (Guidelines for Clinical Effectiveness) for managing vaso-oclusive pain in patients with Sickle cell disease.

2.0 BACKGROUND AND SIGNIFICANCE

2.1 SICKLE CELL DISEASE

Sickle cell disease (SCD) is a term used to describe group of inherited conditions that cause a change in the structure or quantity of the hemoglobin molecule. Hemoglobin is a tetramer comprised of two alpha globin subunits and two beta globin subunits. Mutations in the *HBB* gene can cause abnormal conformation of the beta globin subunit, or beta chain. In particular, a point mutation at the second nucleotide of the sixth codon of the beta chain will cause an amino acid substitution of valine for glutamic acid. This mutation causes an abnormal structural variant of hemoglobin referred to as Hb S. When an individual is homozygous for this particular point mutation, they have Sickle cell anemia (Hb SS). Sickle cell anemia is the most common and typically the most severe form of Sickle cell disease. Other mutations in the *HBB* gene lead to other abnormal versions of beta hemoglobin. These include hemoglobin C and hemoglobin E. Mutations that cause a decrease in the amount of beta globin produced are called beta thalassemias. Compound heterozygosity of Hb S with other atypical forms of hemoglobin can cause Sickle cell disease variations such as SC disease, SE disease, or Sickle/Beta thalassemia. (Wilson, Krishnamurti et al. 2003) In this document the term Sickle cell disease will refer to any variation of Sickle cell disease. All forms of Sickle cell disease (SCD) are autosomal recessive hemoglobinopathies that are characterized by chronic hemolytic anemia and vascular occlusion.

Mutations in the *HBB* gene causing SCD are most often seen in individuals with African, Mediterranean, Middle Eastern, Indian, Caribbean, and Central and South American ancestry although the condition can be found in individuals with any ethnic background. Among African Americans, SCD is diagnosed in approximately 2000 live births annually. This translates to about 1 in 250-600 African Americans with the condition. (Shafer, Lorey et al. 1996)

2.1.1 Pathophysiology of Sickle Cell Disease

The primary function of hemoglobin is to transport oxygen molecules from the lungs to the rest of the body. In individuals with SCD this function is hindered due to the effects of vascular occlusion and hemolytic anemia. Vascular occlusion is associated with the hallmark symptom of SCD: the acute painful episode. The process of vaso-occlusion is complex, but may be broadly described by two events; the polymerization of the hemoglobin S molecule within erythrocytes and adhesion of red blood cells to the endothelium of blood vessels. (Hebbel 2000) When red blood cells that contain hemoglobin S become deoxygenated valine residues are able to "dock" to neighboring beta chains causing the formation of an irregular, "sickled" shape. (Stuart and Nagel 2004) Consequently, the survival of a red blood cell may be reduced from 120 to as low as 20 days. (Wilson, Krishnamurti et al. 2003) Polymerization also affects the deformability of red blood cells. The other key process that occurs in concert with polymerization is adhesion of red cells to the endothelium of blood vessels. Adhesion occurs due to a response of the endothelium to various signals such as hypoxia, thrombin, and platelet activating factor or interleukin-1. (Hebbel 2000) Adherence of reticulocytes and dense, poorly deformable red cells

to the epithelium will impede blood flow, increasing transit time. This in turn causes local anemia from increased red cell turnover. (Stuart and Nagel 2004)

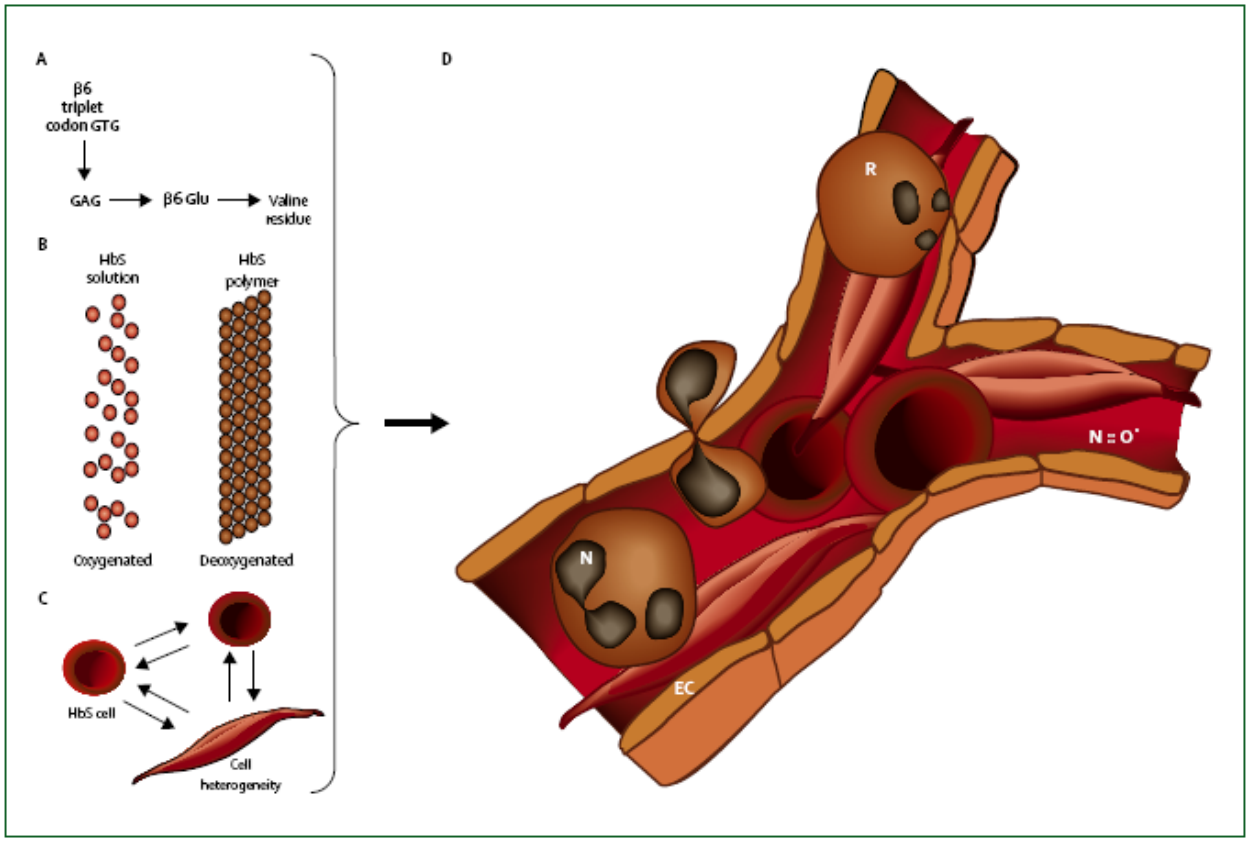


Figure 1. Pathophysiology of Vaso-occlusion. (Stuart and Nagel 2004)

2.1.2 Clinical Features of Sickle Cell Disease

Sickle cell disease is a chronic condition that is associated with a myriad of symptoms that stem from vascular occlusion and hemolytic anemia. Hemolysis can cause anemia, jaundice, pulmonary complications, cholelithiasis, delayed growth and sexual maturation. Vascular occlusion causing tissue ischemia can result in organ damage and dysfunction.

Painful episodes in SCD are complex and can manifest as acute or chronic events. Pain symptoms may be the result of vascular occlusion, secondary to other symptoms of SCD, like leg ulcers or priapism, or result from various treatments of the condition that cause withdrawal symptoms or post-operative pain. (Ballas 2005) However, the primary clinical feature of sickle cell disease is the acute, vaso-occlusive painful episode that requires the use of opioids for alleviation of pain sensation. Vascular occlusion can occur anywhere in the body, likewise pain sensation can be felt anywhere over the body. The most common areas involved in pain events include the chest, back, abdomen and limbs. Dactylitis, or "hand-foot syndrome" is often the first clinical sign of SCD. It presents as a painful swelling of the hands and feet due to vaso-occlusion of the vasculature in this area and may predict a more severe disease course. (Miller, Sleeper et al. 2000) Effective pain management in childhood shapes a patient's coping mechanisms as they get older. As a result, pain should be assessed from a multidisciplinary approach when possible, taking into consideration the life experiences of each patient. (Ballas 2005)

Infection is linked to many complications associated with SCD. Despite preventative measures such as prophylactic penicillin regimens and pneumococcal vaccination, sepsis remains a major concern for patients with SCD and is a common cause of death. (National Institutes of Health 2002) Susceptibility occurs due to splenic infarcts that cause malfunction. Prophylactic penicillin administered daily to children with SCD has shown to reduce the cases of Streptococcus pneumoniae sepsis by 80%. (Gaston, Verter et al. 1986) For children who do not have a history of pneumococcal infection or splenectomy daily, prophylactic penicillin can be stopped at age 5. In addition to all routine vaccinations including the 7-valent pneumococcal conjugate vaccine (Prevnar), children with SCD should also receive the 23-valent polysaccharide

pneumococcal vaccine (PPV). Individuals with SCD should also receive annual influenza and semi-annual meningococcal vaccination. (Mehta, Afenyi-Annan et al. 2006) Due to the increased risk for sepsis, febrile SCD patients with fever of 38.5 degrees Celsius or greater are is an emergency situation. Patients should be evaluated by the Emergency Department, have blood cultures drawn immediately, followed by administration of IV antibiotics. Evaluation should include a physical exam, chest x-ray and blood counts.

The risk for stroke is a major complication with SCD, particularly for individuals with SCD-SS (Sickle cell anemia). Neurological complications may include cerebral infarcts or intracranial hemorrhage. Cerebral infarcts occur most commonly in children with the first recognized cerebral event occurring between the ages of 2-5. (Wong and Powars 2005) Seventeen percent of children with SCD who do not have any neurological symptoms have changes that can be detected on magnetic resonance imaging. (Mehta, Afenyi-Annan et al. 2006) These changes may be associated with learning and behavioral difficulties and may indicate an increased risk for more severe brain ischemia in the future. Lifetime risk for SCD patients for a cerebral infarction is 30%. (Wong and Powars 2005) Strokes occur due to relentless polymerization of Hb S and endothelial changes that lead to a narrowing of the vasculature. Annual screening using transcranial Doppler ultrasonography is recommended for patients with SCD-SS beginning at age 2. Increased blood flow velocity of 200 cm/s or greater on transcranial Doppler indicates significant stenosis and therefore an increased risk for stroke. Transfusion therapy should be considered for these patients. (Wong and Powars 2005)

Pulmonary complications including acute chest syndrome and pulmonary hypertension are major life-threatening concerns for patients with SCD. Pulmonary problems not directly caused by vaso-occlusion, like pneumonia or asthma can exacerbate SCD due to local or

systemic hypoxia that increases polymerization of Hb S, leading to a complex cycle of health concerns.

Diagnosis of acute chest syndrome (ACS) is made in the presence of a new pulmonary infiltrate on a chest x ray with chest or back pain, a temperature of more than 38.5 degrees Celsius, tachypnea, wheezing or cough. (Vichinsky, Neumayr et al. 2000) ACS is the second most common cause for hospitalization and the leading cause of morbidity and mortality in patients with SCD. The cause of ACS is multifactorial, but is commonly associated with pulmonary infarcts due to fat embolism from bone marrow infarcts. Infectious agents, including Streptococcus pneumoniae and parvovirus B19, are also associated with ACS. The strongest risk factor for development of ACS is hemoglobin genotype. Patients with HbSS have the highest incidence, while patients with sickle/beta thalassemia (HbS/β+) have the lowest incidence. Treatment recommendations for ACS include administration of oxygen, blood transfusion, antibiotics, pain control and spirometry. Assessment of blood oxygen requires comparison of baseline arterial blood gases to current arterial blood gases to estimate the alveolar-arterial gradient. Repeated monitoring of the A-a gradient is a good predictor of clinical severity of ACS. Bronchodilator therapy may also be helpful to increase oxygenation for some patients. Exchange transfusion will decrease the proportion of sickle red cells and increase the oxygen affinity of blood. Intravenous broad-spectrum antibiotics should be given upon diagnosis of ACS or febrile patients due to difficulty in excluding bacterial pneumonia. Incentive spirometry can prevent hypoventilation in individuals with ACS and can reduce the development of ACS in individuals with vaso-occlusive pain. (Vichinsky, Neumayr et al. 2000)

It has been shown that 32% of adult SCD patients exhibit pulmonary arterial hypertension, a complex and life-threatening complication of SCD. (Gladwin, Sachdev et al.

2004) Children also exhibit pulmonary hypertension. Although there is not enough data to support overall incidence in the pediatric population, at least one study as has show a correlation with low hemoglobin, elevated reticulocyte count, and cerebral vasculopathy. (Ambrusko, Gunawardena et al. 2006) The underlying pathophysiology of pulmonary hypertension remains to be completely elucidated although it is likely multifactorial. Hb S polymerization decreases deformability and increases fragility of erythrocytes causing their premature breakdown yielding hemolysis. Hemolysis interferes with nitric oxide (NO) synthesis and utilization, which causes dysfunction of the vasculature including NO resistance, initiation of platelet and tissue factors, vasoconstriction, and vaso-proliferation. (Castro and Gladwin 2005) Chronic hemolysis is a risk factor for pulmonary hypertension. As a result, individuals with HbSS genotype are more likely to acquire pulmonary hypertension than someone with Hb SC genotype. Screening involves the use of a transthoracic Doppler echocardiogram that measures tricuspid regurgitant jet velocity (TRV). A TRV value of 2.5m/sec or greater is considered a clinically significant elevation suggesting an increased risk for morbidity and mortality from pulmonary hypertension. (Ambrusko, Gunawardena et al. 2006) Pulmonary arterial pressures of 5mmHg and 15mmHg are estimated to have vaso-constriction of greater than 50% and less than 50%, respectively. In one study the median survival with pulmonary hypertension (25mmHg) was 2.8 years. (Castro and Gladwin 2005) Management and treatment of pulmonary hypertension are still being determined but will likely involve Hydroxyurea therapy, exchange transfusion, iron chelation therapy, and vasodilators dilators including prostacyclins. (Gladwin, Sachdev et al. 2004; Castro and Gladwin 2005)

Acute splenic sequestration is a relatively common and potentially life-threatening condition caused by accumulation irregular red cells by the spleen. (Aquino, Norvell et al. 1997)

Confinement of blood in the splenic sinusoids causes splenomegaly, a significant drop in hemoglobin level, and the potential for hypoxic shock. Acute splenic sequestration typically occurs between ages 3 months and 5 years, but can occur at any age. Individuals with Hb SS typically present with acute splenic sequestration early in childhood, while those with Hb SC typically present with symptoms later in childhood. Patients and caretakers should be informed of the importance and method of palpating the spleen. (Aquino, Norvell et al. 1997) Splenic sequestration can be diagnosed with an acute decrease of 2g/dl of hemoglobin from average baseline hemoglobin, elevated reticulocyte count as compared to baseline counts, and enlarged spleen greater than baseline measurements. Clinical symptoms associated with splenic sequestration may include: weakness, pallor, and tachycardia. Treatment is aimed at restoring circulating blood volume with transfusion. Due to the high rate of recurrence, spleen management in young children often involves chronic transfusion and/or splenectomy. Issues to consider when making decisions regarding management should include the risk for infection following splenectomy, consideration of whether chronic transfusions will restore splenic function, and ability of the child's caregivers to identify, react, or access care during subsequent splenic sequestration.

Other major health complications for individuals with SCD include: dysfunction of biliary tract and liver, renal changes, priapism, leg ulcers, and bone complications. Gall bladder and liver problems likely stem from chronic hemolysis. This causes an increase in bilirubin production and incidence of gall stones. Almost 30% of patients with SCD develop gall stones by age 18. Treatment involves the use of antibiotics with the option of cholecystectomy. Liver involvement may be caused by vascular occlusion or chronic hepatitis. Exchange transfusion is the most commonly used treatment option.

The kidney is especially at risk for organ damage from Hb S polymerization and vaso-occlusion due to the local environment that is hypoxic, acidic, and hypertonic. Hyposthenuria is the most common renal complication in SCD. As a result these individuals are also at increased risk for dehydration, which is a risk factor for vaso-occlusive pain event. Patients should be encouraged to always stay hydrated. Other changes in the kidney result in proteinuria and hematuria. Both conditions overtime can be life-threatening, treatment for some patients may ultimately involve kidney transplant. (Ataga and Orringer 2000)

Priapism in males with SCD is caused by vaso-occlusion that causes obstruction of the venous drainage of the penis. Priapism is a common complication for males with SCD, with 89% of males experiencing at least one episode by age 20. (Mantadakis, Cavender et al. 1999) Treatment may include IV hydration, analgesics, or penile aspiration of blood.

Bone marrow hyperplasia from chronic hemolytic anemia, vaso-occlusive episodes causing bone infarcts, and sepsis resulting in osteomyelitis are the primary causes of bone complications for patients with SCD and are a mail cause of morbidity. Another common complication for individuals with SCD is the occurrence of leg ulcers. Although their etiology is unclear, they occur in 10-20% of patients with SCD.

2.2 PAIN MANAGEMENT IN THE EMERGENCY DEPARTMENT

Despite published guidelines for pain management by organizations such as JCAHO, and ample availability of effective pain medication and other therapies in health care settings, undertreatment, or oligoanalgesia, in emergency medicine continues to be a major public health

problem. (Berry and Dahl 2000; Phillips 2000; Rupp and Delaney 2004) Inadequate pain management in hospital settings was brought to the forefront following a 1973 article by psychiatrists Marks and Sachar. (Marks RM 1973) Since that time a plethora of literature has been produced stating similar findings that poor pain management continues to be recognized, and continues to be inadequate. Pain that continues to be ineffectively managed has physiological and psychological effects directly impacting overall quality of life. (Ballas, Barton et al. 2006) Chronic pain can impact family dynamics, social relationships, and employment status. Inadequate pain relief creates a financial burden on the health care system and on society costing millions of dollars annually as a result of longer hospital stays, re-hospitalizations, and emergency department visits. Individuals who are unable to work may lose income as well as insurance coverage, further perpetuating the public health dilemma. Although efforts by the Joint Commission on Accreditation of Healthcare Organizations (JCAHO) and other accrediting organizations to create standardized treatment guideline for managing pain in the Emergency Department have been established, recent literature describes little improvement. (Todd KH 2007)

The acute pain episode is the most frequent complication in patients with SCD and is associated with diminished quality of life. (Benjamin, Swinson et al. 2000) Pain episodes are unpredicable, notoriously intense, and continue to recur over a lifetime. Effective pain management for patients presenting to the ED with SCD and other chronic illnesses continues to be a struggle in many institutions. There are several proposed reasons for this. One reason is that health care providers have preconceived ideas regarding patients seeking analgesia, or inappropriate concern for causing addiction in patients. (Rupp and Delaney 2004) It has also been proposed that poor pain management may stem from a lack of emphasis in medical school

curriculums or residency programs. (Augarten A 2006) Another explanation is the absence of tangible proof of pain in conditions like SCD may lead physicians to under-appreciate the magnitude of the pain episode. (Elander J 2006) Additionally, there is general lack of understanding of SCD and consequently poor appreciation of how to effectively manage pain in patients with chronic exposure to opioids. (Benjamin, Swinson et al. 2000)

2.2.1 Opioid use in pain management

Opioid agonists are commonly used in management of pain episodes for patients with SCD. Opioid agonists achieve pain reduction by decreasing the perception of pain by the central nervous system. Adverse effects of this class of narcotics include itching, nausea, vomiting, sedation, seizures and respiratory depression. Opioids have no ceiling effect, where eventually doses no longer have an additive effect. As a result, the only reason to limit opioid administration in a patient is concern for the adverse effects listed above. (Ballas 2005)

2.2.2 Pain Management Acute Vaso-occlusive Pain Episode and Discriminating Between Analgesic Dependence, Tolerance, Addiction and Psuedoaddiction

In one of her numerous articles published in 1980 Margo McCaffery, RN, MS said, "Pain is whatever the experiencing person says it is, existing whenever he says it does." (McCaffery 1980) This simple and elegant statement captures the subjective nature of pain sensation nicely. Inappropriate misconceptions regarding drug addiction may account for many cases of poor management of pain. Analgesic dependence and tolerance occur after chronic use of a drug

causes physiological changes. Tolerance occurs when a particular dose loses its effectiveness and desired effects can only be achieved with larger doses. Dependence occurs when a physiological change occurs in the body where withdrawal symptoms, such as sweating, nausea, diarrhea, vomiting or seizures occur when the drug is no longer taken. Genuine analgesic addition among patients complaining of pain is rare. Addiction may occur with or without physiological responses. Addiction is the compulsive desire to take analgesics for effects other than pain relief. It is differentiated from dependence in that individuals exhibit behavior that indicates a compulsive need to have the drug. (McCaffery and Hart 1976; Shapiro BS 1994) Pseudoaddiction occurs when a patient historically has had poorly managed pain in the health-care setting. These individuals resort to behaviors that resemble drug-seeking behavior and are interpreted as such by health-care providers. This cycle of poorly managed pain and manipulative behavior by patients in a pain episode undermine trust between groups, further compromising effective pain management. (Lusher, Elander et al. 2006)

Health care providers who treat pain are influenced by contextual factors that surround the interaction between patient and provider. Inaccurate perception and overestimation of addiction may be attributed because of the resemblance of certain pain behaviors that resemble symptoms of analgesic addiction. This in turn can lead to increased anxiety during visits to the emeregency department and displays of pseudoaddiction in patients with SCD. Patients must resort to exaggerated or manipulative behavior in order to persuade health care providers to administer appropriate analgesics. (Elander J 2006) Pain episodes in SCD are unique in that they are recurrent over a lifetime and typically don't present with objective, verifiable symptom. Most conditions that exhibit pain occur as a one time event, either acutely (following surgery) or chronically (as in cancer). Patients who experience pain episodes as one time events do not have

the opportunity to learn coping mechanisms as patient with SCD do, that can cause another layer of complexity for health-care providers who must differentiate between dependence and abuse (Lusher, Elander et al. 2006).

In 2006, Elander, et al. published a vignette study where health care providers were asked to examine fictious patients with SCD who displayed genuine symptoms of analgesic addiction and pain behaviors that resemble those symptoms. Providers judged pain level, likelihood of addiction, and the analgesic needs. The results showed that providers were able to distinguish between true addictive behavior and behaviors that resemble addiction. However, no differences were seen in the decisions regarding amount of required analgesia needed for both sets of patients. Health care providers treated patients they correctly identified with behaviors only resembling addictive behavior (pseudoaddiction), with less powerful or less frequent analgesics. Health-care providers who practice oligoanalgesia will further perpetuate anxiety and behaviors of pseudoaddiction in patients. Health care providers (nurses and doctors) with more experience in treating patients with SCD made higher estimates of analgesic needs indicating that a deeper understanding and appreciation for the condition yields more rigorous treatment of pain in patients with SCD. Elander concluded that staff training should emphasize less on formal knowledge of SCD and more on practical experience in making assessments of patients who require analgesia. (Elander J 2006)

2.2.3 Hydroxyurea

Hydroxyurea is a chemotherapeutic drug that can reduce the number to SCD-related complications including acute chest syndrome, and lowering number of pain episodes by 50%.

(Ballas, Barton et al. 2006) The exact mechanism of how hydroxyurea works is unclear however effects on the bone marrow include an increase in the synthesis of fetal hemoglobin (Hb F). (Charache, Terrin et al. 1995) Historically, it was the observation of patients with persistently high HbF levels and milder disease course that lead to the addition of hydroxyurea as a treatment option. This makes sense as Hb F does not have a beta globin subunit, it is a tetramer comprised of to alpha and two gamma globin chains. Due to the cytotoxic nature of this drug, it is unclear what the long-term effects will be. With that in mind, hydroxyurea is prescribed to pediatric patients who present with a more severe disease course.

2.2.4 Pain Management and Effect on Quality of Life

"…pain is not equal to or reducible to vao-occlusion. Pain is a multifaceted, complex, and inherently subjective experience. The physiologic process of vaso-occlusion may initiate and maintain the experience of pain, but the pain cannot be reduced to the pathophysiology." (Shapiro B 1993) To provide effective manangement of pain in SCD, health care providers must have complete understanding of the issues associated with the treatment of pain of an incurable disease on a chronic basis. Pain in SCD is not unlike other types of pain where it is a complex human experience. Strongly influenced by social, cultural and spiritual experiences. As a result, pain management becomes more effective as health care providers learn to know their patients beyond the initial triage questions. (Ballas 2005)

Health-related quality of life is a layered concept that includes physical, emotional, and psychosocial pieces associated with a disease or treatment. Poor pain management in the emergency department (ED) can impact all areas of life for individuals with Sickle cell disease.

(Ballas 2005) Maintaining a high quality of life in patients with a chronic disese and chronic pain is difficult. Individuals who live with recurrent, acute pain episodes are known to have increased risk for mental health problems, lower performance at work, and general lower quality of life. (Ballas, Barton et al. 2006) Additionally, children with SCD exhibit lower performance at school and impaired social interaction. The chronic, lifelong nature of SCD and it's impact on all areas of life can be best treated with a multidisiplinary approach by providers that know their SCD well, addressing phyical, emotional and psycosocial needs. (Fuggle, Shand et al. 1996; Benjamin, Swinson et al. 2000)

Although the understanding of the pathophysiology of SCD continues to become more and more clear, and many advances are achieved in availability of treamtent and management options to alleviate symptoms, basic principles of pain mangement are not employed by many sickle cell experts. Clinical features are not well understood by pain specialists, and many physicians and nurses lack general basic knowledge of SCD and pain management. It is well recognized that ineffective pain management increases morbidity. Furthermore, there is an association with number of pain episodes per year and mortality. (Benjamin, Swinson et al. 2000) Despite efforts by JCAHO to establish pain assessment guidelines and make pain the "fifth vital sign" to increase awareness by health care providers that pain is an important and undertreated symptom, changes in intervention or improvements in treating pain has not been acomplished by all institutions. (Narasimhaswamy, Vedi et al. 2006) New methods must be explored to execute changes in practice by health care providers treating pain in SCD.

2.3 WEB-BASED LEARNING

As medical education continues to expand into alternative delivery methods, web-based learning has emerged as a teaching opportunity to make education more accessible and more relevant to learners. Web-based learning provides several advantages. It provides an efficient and convienient method to reach many health-care providers. The vast availability of personal computers, faster Internet connections, and the use of the World Wide Web and other electronic resources as instant references, makes web-based learning a seamless addition to traditional teaching methods. Additionally, web-based education would allow some learners to gain insight into topic areas that they do not have much practical experience, and allow more seasoned providers an opportunity to learn more about the latest advances in treatment—all with instantaneous access.

Compared to traditional teaching, web-based learning provides an opportunity to review complex topics using an interactive method that provides links to relevant literature, and concise didactic segments that cover the most essential information. Typically, learners will access educational modules when it is convenient for them, allowing for a more relaxed, and thorough learning experience.

2.3.1 The Virtual Patient

Professional development of health-care providers is achieved by quality learning, training, and problem-solving. Often much of this development occurs as providers begin to interact with patients. Unfortunately, for a variety of reasons this interaction is limited. Out-patient care,

ethical considerations, unwillingness of patients, and scarcity of certain types of patients prevent continued development of problem-solving skills, and training. One way to provide learners an opportunity for increased experience is by creating a virtual patient experience.

Virtual patients can use a problem-solving approach where the learner must gather information from physical exam, labs, or medical history and then diagnose or manage the patient. Virtual patients may also have a narrative approach that follows a storyline. Virtual patients allow for repetitive, non-judgmental practice of clinical skills by the learner. This provides an environment where mistakes are allowed, and safety is maintained for patients and providers. A web-based platform is an effective medium to utilize virtual patient cases. Changes and refinements can be continually addressed, more than one learner can access the application at one time, and applications can be access at any time. Additionally, the application is installed onto a server to that learners do not have to bother with software components, and finally cases may be utilized by an infinite amount of individuals, allowing for wide distribution of information.

2.3.2 Learning Styles and Web-based Learning

It is well established that there are a variety of learning styles that influence the way that people learn and interpret information. If teaching methods are different from learning style, this could negatively affect the learners' performance. (Curry 1999) As web-based educational applications reach large, heterogeneous populations, the variety of learning styles could potentially have an impact on effectiveness and adaptation of the application by learners. In

2005 David Cook, MD published an article that reviewed various styles of learning and considered their relationship to web-based education applications. (Cook 2005)

The wholist-analytic dimension described by Cook is derived from various studies that all classify learning styles. This dimension describes how these individuals interpret various situations to understand the educational message. They see a unified whole, or as a collection of individual parts. Wholists are guided by a broader frame of reference while analytics extract clues from the overall picture that allow for step-wise learning. In terms of web-based education, analytics appear to perform better when there is less structure, and there is encouragement for detailed investigation before garnering the overall message. Wholists perform better when there is some structure and direction through an educational application. Additionally, wholists may benefit from more discussion based interaction, where there is more social interaction with other learners. (Cook 2005)

The verbalizer-imager dimension encompasses several descriptions of learning and cognitive style. Cognitive style describes personal attributes like perception, memory, and judgment that be related to educational contexts and non-educational contexts. Verbalizers, as the name insinuates, learn best from speech and written text. Socially, verbalizers will be better communicators and will likely only tolerate stimulating presentation. Imagers learn best from pictures and demonstration. They are described as better than verbalizers at keeping track of their location in space, which could translate into better performance in a complex, web-based application. Imagers are further described as more likely to tolerate tasks they find unappealing. On the surface it would seem that verbalizers would perform better in web-based applications that were text-heavy and more directed, while imagers would do well with more multi-media and hyperlinks, without being distracted by a potentially convoluted path through cyberspace. The

evidence however among medical education, is mixed. There is no significant identified difference in scores between these groups. Evidence could be confounded if there are a higher proportion of certain types of learners in the health-care field.

The concrete-abstract dimension describes how information is acquired and then processed. Knowledge acquisition is either concrete though the senses, or abstract by analyzing. The concrete learner prefers learning from experience and example, while the abstract learner prefers concepts and theory. Preferred instructional method for the concrete learner would involve case-based learning with interactive demonstration. The use of factual information would favor abstract learners. Limited evidence has not demonstrated difference in performance by these two learning styles. Adaptation in instructional method may provide more impact on learning than changes to presentation or environment of a web-based program.

The active-reflective dimension describes how learners use the knowledge they have gained. Active learners are more apt to utilize the information without consideration of the implications. Reflective learners tend to observe, internalize, and examine different perspectives as they assimilate the information. Due to the independent and self-paced nature of web-based education, reflective learners would seem to enjoy and perform better with this type of learning. The exception to this would be if an application required several interactive features that would disrupt the reflective learners need for time to process the information. Interactive features would however suit the needs for an active learner. Limited evidence available indicates that active learners do indeed perform better than reflective learners in web-based learning formats.

There is literature to support that different types of learners perform better when teaching methods are adapted to specific learning style. Information is limited due to the dearth of research in this area, small sample sizes, and differing educational applications. Cook

recommends that future web-based studies measure variation in design, presentation and teaching method, rather than focusing on whether learners perform better tradition or computer-based methods. (Cook 2005)

2.3.3 Educational Intervention on Pain Management in the Emergency Department

Patients presenting to the ED with pain typically have some level of anxiety that is either produced by the underlying cause for the pain or due to past experiences in the health care setting. Pain is historically undertreated, this causes increased anxiety for patients with SCD who live with a chronic condition that causes recurrent pain episodes over a lifetime. In a study performed by a group in Israel, pain management in the ED improved substantially following educational interventions. The most significant improvement in management was achieved following a one day course in pain management that included a lecture and patient simulation with actors. (Augarten A 2006)

Health care provides in the Emergency department (ED) are familiar with a fast-paced, unpredictable, need-to-know working environment. This is inherently problematic when providers are then asked to implement a change in their practice, or to take time to participate in continuing education. Shift work hours, and unpredictable patient flow can create barriers to traditional teaching methods. Web-based educational applications can provide an effective alternative for enhancement or continuing education for health care providers in the ED. Health care workers had an increased response to a web-based learning experience compared to face to face workshops or lectures. Although workshops were designed based on requests of workers, attendance was often low. The ability for individuals to access the educational application at a

time that was convenient for them was likely a contributing factor. (Curran-Smith and Best 2004)

3.0 MATERIALS AND METHODS

3.1 PROJECT DEVELOPMENT

This project has come to fruition as a result of collaboration between the Comprehensive Hemoglobinopathy Program at Children's Hospital of Pittsburgh (CHP) and the Laboratory for Educational Technology (LET) at University of Pittsburgh School of Medicine. Following meetings and discussion to mold the desired educational product, the first major step of was to develop learning objectives for the educational module. The objectives provide an outline of content to be included in the case, and were derived from CHP's Guidelines for Clinical Effectiveness entitled, "Sickle Cell Disease Vasocclusive Crisis in the ED Pain Management". The guidelines are structured as an algorithm to provide a step-wise method for treating pain in patients with Sickle cell disease. The guidelines were created by a group of Sickle cell experts including Dr. Lakshmanan Krishnamurti, Director of the Comprehensive Hemoglobinopathy Program and other team members of the Comprehensive Hemoglobinopathies Program at CHP.

The next step was writing and developing the case that would serve as the foundation of our educational module. The case of a young boy presenting the Emergency department (ED) was written to be a succinct, accurate, typical presentation of a patient with SCD presenting to the ED with pain. Additionally, the case is written to be appropriate for any health-care provider who cares for patients with SCD; physicians, nurses, social workers, and so forth. Users of the

educational module are informed they are a pediatric Emergency Medicine resident. The storyline involves a young boy who presents to the ED with increasing pain in his legs. As they progress through the case, initially the boy's pain is being controlled. However, he suddenly develops a fever and pain symptoms return in another area of the body. The boy is eventually diagnosed with acute chest syndrome and is admitted to the hospital. There he is placed on a PCA (patient-controlled analgesia) and discharged several days later. Interspersed through the case there are several multiple choice questions placed when decisions must be made by the learner regarding management of the patient's pain. Learners can answer the questions as many times as they want and comments are provided as to why an answer was wrong or right. Additionally there are two didactic segments, one about pain scales and the other describes co-morbid conditions that often occur with pain episodes in patients with SCD.

There were several individuals who were consulted to create a case that would be appropriate, realistic and useful for this type of educational intervention. Dr. Krishnamurti, Allison Sakara, CRNP, and Elizabeth Gettig, MS, CGC provided invaluable direction and editing expertise. Dr. James McGee, Director of LET provided essential insight and clarity about structure, content and design of educational modalities that make this project work.

3.1.1 Measuring utility and opinion

To measure the utility of this educational tool, an evaluation was created to gather information (Appendix E). This information will be invaluable to further refine the focus and style of this module, and potentially for similar modules in the future. The evaluation consists of two demographic questions and nine opinion questions. Demographic information will help

determine the population of subjects who are choosing to participate in this activity and also estimate level of experience in treating patients with SCD. There are nine opinion questions that ask learners various questions about their attitudes regarding content and style of information presented in the module. Answer choices to eight of nine opinion questions follow a Likert-type scale with options ranging between "very low" and "very high". The last opinion question asks how the learner will alter their management for patients with SCD. Answer choices range from not changing management at all to changing management immediately. Additionally, there is a page for free-text where learners are encouraged to give feedback, insight and comments.

3.1.2 Measuring Knowledge Gained

To measure knowledge gained by learners participating in this educational activity, 5 multiple choice questions are administered as a pre-test, prior the going through the case, and then as a post-test, following completion of the case. These questions were written as five key ideas that the learner should walk away with following the completion of this module. Questions ask about initial evaluation and opioid dosing for a SCD patient presenting to the ED with pain, primary goal of pain management in SCD, and assessment and intervention methods. (Appendix F)

3.1.3 IRB Approval

This project was submitted for IRB exempt research approval (Appendix B). IRB approval number 0702021 was granted, allowing for collection of responses from the pre-test, post-test and evaluation. Prior to each question set, learners were informed of our overall goals with this

study, and were informed that all information will be kept anonymous and collected for review. Additionally, learners are informed of the option to skip the question sets.

3.2 HOSTING OF THE EDUCATIONAL MODULE

The Laboratory of Educational Technology (LET) is part of the University of Pittsburgh School of Medicine. As medical education increasingly becomes supplemented and enhanced with web-based technology, the LET was established to address the needs for development of this technology. The LET works directly with the Curriculum Committee, the Office of Medical Education, students and faculty of the University of Pittsburgh to create new approaches to learning, including computer and web-based solutions. Dr. James McGee, Director of the LET worked with his group to develop a series of web-based initiatives. Educators and students can access a variety of on-line tools to enhance and supplement traditional classroom learning or increase collaboration between student researchers and advisors.

One of the LET's suite of initiatives includes a collection of web-based virtual patient cases. Here we have posted our educational module entitled, "Pain Management in Sickle Cell Disease". The link to access this module is posted on the Sickle Cell Program web page on the CHP's web site, http://www.chp.edu/centers/03_hema_sickle.php and on PedsEd, an on-line resource for health-care providers at CHP, http://pedsed.pitt.edu/06_browse.asp. PedsEd is also supported by individuals in the LET. Both locations allow for easy access by learners. Sample screen shots of the module are included in this document in Appendix A.

3.2.1 Data Collection

For users that access the case via CHP's Sickle Cell Program web page, the module tallies answer selection by users and creates a detailed report with the information summarized. With a log-in and password, access to a detailed report was available at any time to continually monitor overall tallies. For users that access the case via CHP's PedsEd site, the module tallies answer selection by users; however, no detailed report is accessible outside the LET. To collect data from users who access the case here, members of the LET group will pull periodic reports.

3.3 RECRUITMENT

This educational module was created with the idea that it would be appropriate for any health care provider. To disseminate and advertise the educational module, Dr. Krishnamurti sent information and description of the project via e-mail to colleagues within UPMC including physicians and faculty in Emergency Medicine and Hematology. Also, emails were sent out to Sickle cell experts outside of Pittsburgh, including clinicians associated with other hospital-based Comprehensive Sickle Cell Programs, and clinicians affiliated with the Sickle Cell Disease Association of America (SCDAA). (Appendix C)

4.0 RESULTS

4.1 DEMOGRAPHICS AND EVALUATION QUESTIONS

The results of the evaluation and demographic questions are shown below. Study subjects gave job title, information about number of years experience and opinions regarding this educational module.

Table 1. Job Title of Subjects

Job Title	Number of Subjects	Percentage of Subjects
Resident	3	23.08%
Fellow	2	15.38%
Hematology Attending	2	15.38%
Emergency Medicine Attending	1	7.69%
Nurse Practitioner/Physician Assistant	2	15.38%
Genetic Counselor/Social Worker	3	23.08%
Registered Nurse	0	0.00%
Psychiatrist/Psychologist	0	0.00%
Total responses	13	

Job title of subjects varied significantly. Almost 40% of subjects were either fellows or attendings. About 15% of subjects were nurse practitioners or physician assistants, 23% were residents, while another 23% were either genetic counselors or social workers. (Table 1).

Table 2. Number of Years Since Highest Degree Acheived

Number of Years Since Highest Degree Achieved	Number of Subjects	Percentage of Subjects
0-3	3	20.00%
4-6	7	46.67%
7-9	1	6.67%
10-14	2	13.33%
15+	2	13.33%
Total responses	15	

About 54% of subjects received their highest degree between 4 and 9 years ago. About 27% received their highest degree 10 or more years ago, and 20% received their highest degree within the last 3 years (Table 2).

Table 3 summarizes the results from Evaluation Questions 1 through 8. Evaluation question 1 shows 35% of subjects felt that the educational objectives were achieved to a "very high" extent. 29% felt they were achieved to a "high" extent, and 35% felt they were achieved to a "moderate" extent. Evaluation question 2 measured subjects' feelings on the quality of the module. 35% were satisfied to a "very high" degree with the quality. 41% felt a "high" degree of satisfaction and 24% were "moderately" satisfied with the quality. Evaluation question 3 asked if users felt this module was relevant to their practice. 81% of subjects felt that content of the educational module was relevant to their practice to a "very high" or "high" degree, and 19% felt content was relevant to a "moderate" degree. Users opinions as to whether they felt a web-based educational module was useful was solicited in evaluation question 4. 67% of subjects felt this type of educational intervention is useful to a "very high" degree.

Table 3. Answer Responses to Evaluation Questions 1-8

Answer Responses % (n)

	Very Low	Low	Moderate	High	Very High	Totals
Evaluation Q1	0	0	35.3% (6)	29.4% (5)	35.3% (6)	(17)
Evaluation Q2	0	0	23.5% (4)	41.2% (7)	35.3% (6)	(17)
Evaluation Q3	0	0	18.8% (3)	43.8% (7)	37.5% (6)	(16)
Evaluation Q4	0	0	20% (3)	13.3% (2)	66.7% (10)	(15)
Evaluation Q5	0	7.7% (1)	53.9% (7)	30.8% (4)	7.7% (1)	(13)
Evaluation Q6	0	50% (6)	25% (3)	8.3% (1)	16.7% (2)	(12)
Evaluation Q7	0	38.5% (5)	53.9% (7)	7.7% (1)	0	(13)
Evaluation Q8	0	0	63.6% (7)	9.1% (1)	27.3% (3)	(11)
Totals	0	(12)	(40)	(28)	(34)	

Evaluation Question 1: To what extent were the educational objectives achieved?
Evaluation Question 2: To what extent were you satisfied with the overall quality of the educational activity?
Evaluation Question 3: To what extent was the content of the educational activity relevant to your practice?
Evaluation Question 4: To what extent do you feel that this type of web-based educational tool is useful?
Evaluation Question 5: To what extent did the activity enhance your knowledge of treating pain in patients with Sickle cell disease?
Evaluation Question 6: To what extent did the activity change the way you think about treating pain in patients with Sickle cell disease?
Evaluation Question 7: To what extent will you make a change in your practice of treating pain in patients with Sickle cell disease as a result of your participation in this educational activity?
Evaluation Question 8: To what extent did the activity help you to learn and understand the algorithm provided (Vasoocclusive Pain in Sickle Cell Disease)?

13% felt the intervention was useful to a "high" degree and 20% felt in was "moderately" useful. Evaluation question 5 asked subjects the degree to which this educational module enhanced their knowledge of pain management in SCD. 8% felt it enhanced their knowledge to a "very high" degree, 31% felt the module enhanced their knowledge to a "high" degree and 54% felt this educational module enhanced their knowledge to a "moderate" degree. 8% felt the module enhanced their knowledge only to a "low" degree. Evaluation question 6 asked to what extent the module changed the way they think about pain management. 17% felt the module changed their thinking to a "very high" degree, 8% felt the change was "high", 25% felt they changed their way of thinking to a moderate degree, and 50% reported that this module changed their way

of thinking about pain management to a "low" degree. The next question, evaluation question 7, asked to what extent users would implement a change in the way they practice pain management in SCD. 8% reported they would change their practice to a "high" degree, 54% felt they would change to a "moderate" degree, and 39% reported they would change their practice to a "low" degree. No one felt they would change their practice to a "very high" degree following the use of this educational module. In evaluation questions 5, 6, and 7 several individuals chose the "low" answer choice. These three questions pertained to how much the content of this educational module enhanced their existing knowledge of pain management to initiate a change in their thinking or practice of pain management. Evaluation question 8 asked whether the educational module helped users to learn and understand the CHP algorithms that were provided. About 27% said the module helped to a "very high" degree, 9% said it helped to a "high" degree and 67% said it helped to a "moderate" degree. For all eight evaluation questions, no responded that the degree to which something was achieved was "very low".

The final evaluation question (Figure 1) asked how learners will change their management of pain in patients with SCD. 73% (8/11 responses) said they "will not change, because they are treating in accordance with the algorithm. 9% (1/11 responses) said they would "need more information before they make a change" in practice, and 18% (2/11 responses) said they would "immediately change" their management of pain in patients with SCD.

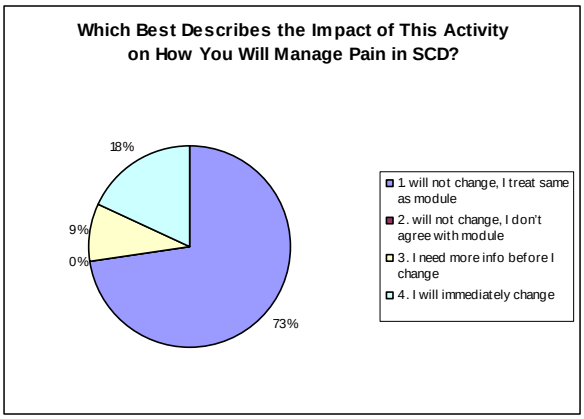


Figure 2. Evaluation Question 9

4.2 KNOWLEDGE QUESTIONS

The knowledge questions were written to determine whether there was a shift understanding of the basic principles of treating a patient with SCD in the ED.

Table 4. Answer Selection to Knowledge Questions

	#correct/ #incorrect (%correct) Pre-Test	*#correct/ #incorrect (%correct)* Post-Test
Knowledge Question 1	12/20 (60%)	7/13 (54%)
Knowledge Question 2	16/19 (84.2%)	12/13 (92.3%)
Knowledge Question 3	16/17 (94.1%)	13/13 (100%)
Knowledge Question 4	7/16 (53.3%)	9/13 (69.2%)
Knowledge Question 5	12/17 (70.6%)	13/13 (100%)

See Appendix F for knowledge questions and answer options.

Knowledge question 1 asks the learner what should be evaluated for when a patient with SCD presents to the ED with pain and fever. The correct response is that complications associated with Sickle cell disease and also other common medical problems should be considered. In the pre-test, answers were given for all choices available with 60% answering the question correctly. 25% felt that only the common co-morbidities should be evaluated for, 10% felt in this scenario only common medical or surgical complications should considered, and 5% thought it likely that this patient was attempting to obtain more pain medication. Interestingly, in the post-test the number of subjects choosing the right answer decreased to 54%, with 46% of subjects choosing evaluation of common complications of Sickle cell disease only as the correct answer.

Knowledge question 2 asks the best way to determine initial dosing of pain medications for patients with SCD. The correct response is to follow the patients established Individualized Management Plan or the institutional guidelines for treating pain in patients with SCD. About 84% of pre-test subjects chose the correct answer. Approximately 5% of pre-test subjects felt that all SCD patients should receive the same management with IV fluids, oxygen and Ketorolac followed by a 2 hour interval to determine if opioids should be considered. Another 5% of pre-test subjects felt the patient should receive what they were taking at home for pain. The remaining 5% felt that dosing should start at the low end of the range for body weight and age. 92% of post-test subjects chose the correct response, with 8% feeling the patient should continue to receive what they were taking at home.

Knowledge question 3 asks learner what they think the overall goal is for treating pain in patients with SCD presenting to the ED with a pain episode and how the role of opioids administration should be approached. The correct answer is “aggressive management of pain in

a supportive environment can lead to relief of pain and obviate the need for hospitalization in a substantial proportion of patients". About 94% of pre-test subjects answered correctly, with the remaining 6% of pre-test subjects feeling that opioids should be restricted for this patient population due to concerns for developing drug dependence and tolerance. 100% of post-test subjects answered this question correctly.

Knowledge question 4 asks what the intervals should be for administering pain medications to a patient with SCD who is experiencing a pain episode. The correct response is, "peak effect of morphine occurs in 20 minutes. Based on patient response, doses can be repeated at that interval". About 56% of pre-test subjects answered correctly. 19% of pre-test subjects thought the correct response was that pain medications can be administered every 3-5 hours, another 19% thought it was dependent on whether the patient was a child or an adult, and about 6% felt all patients with SCD should receive fixed doses of Demerol. There was an increase to 69% giving the correct response in the post-test. Additionally, 8% still felt that intervals were every 3-5 hours, and 23% thought it was dependent on whether it was a pediatric or adult patient.

Knowledge question 5 asks about the best way to assess the patient's response to current management of pain. About 71% of pre-test subjects answered correctly, feeling the best way to make this determination is by self-report by the patient. 12% thought the best way would be through nursing staff observation of patient activity, and 18% thought consideration of clinical assessment, vital signs and laboratory markers for inflammation was the most appropriate way. 100% of post-test subjects answered this question correctly.

5.0 DISCUSSION

5.1 DEMOGRAPHICS

Demographic information obtained in this project was limited to job title and number of years since highest degree was achieved. Collecting this information roughly determines the amount of past experience of our study population. In both measures, the study population is quite varied. This is advantageous for patients in that increased knowledge of effective pain management by all health-care providers who care for patients with SCD will yield better overall care. A varied study population can also have an impact on degree of knowledge gained. Understandably, an Emergency Medicine attending is going to have more experience in pain management and therefore higher preliminary knowledge of pain management than a social worker. On the other hand, individuals who have been practicing for many years may be more reluctant to change their current practices. For example, if a physician has been under the assumption that patients with pain should be reassessed at the medication's estimated half-life for the last 10 years, it may take more effort to convince them to begin to reassess at the medication's peak effect instead. It may be easier to influence practice among health-care providers who are earlier in their career and still developing their skill sets.

Recruitment for subjects to participate involved distribution of an email describing our efforts and asking for recipient participation as well as asking recipients to recruit their

colleagues to also participate in our educational module. Email recipients included Hematology and Emergency Medicine faculty within University of Pittsburgh Medical Center (UPMC) and also to Sickle cell experts outside of Pittsburgh, including clinicians associated with other hospital-based Comprehensive Sickle Cell Programs, and clinicians affiliated with the Sickle Cell Disease Association of America (SCDAA). While this population of individuals likely already has strong understanding of effective pain management, they provide valuable feedback to help future refinement of the module.

5.2 AIM 1: ASSESS WHETHER HEALTH CARE PROVIDERS HAVE INCREASED KNOWLEDGE OF MANAGEMENT ACUTE PAIN FROM VASO-OCCLUSION IN PATIENTS WITH SCD

To determine whether this objective was achieved, evaluation question 5, "To what extent did the activity enhance your knowledge of treating pain in patients with Sickle cell disease?" and answer selection from the pre and post test knowledge questions were considered.

Answers from evaluation question 5 ranged from "low" enhacement of knowledge to a "very high" degree of enhancement of knowledge, with the vast majority (84%) feeling knowledge enhancement was to a "moderate" or "high" degree. This variability in responses likely reflects the variety of background knowledge that our subjects have. Although it is somewhat reassuring that subjects feel that they have some enhancement of knowledge, results from the pre and post test indicate there are some individuals who have not incorporated some of the most important teaching points. This indicates that refinement in our educational module is required.

Additionally, not surprisingly, it is clear that there is a high level of preliminary knowledge in this subject population. As a result, there is not a sharp shift in knowledge gained. Assessment of knowledge gained is also limited by a small sample size and differing sample size from pre-tests (about 20) to post-tests (13).

There was general improvement number of correct responses from the pre-test to the post-test in 4 out of 5 knowledge questions. Surprisingly there were fewer correct responses on the post-test than on the pre-test for Knowledge Question 1, "A patient with SCD presenting with fever and chest or abdominal pain should be evaluated for which of the following?" The correct response is that the patient should be evaluated for the common co-morbidities associated with SCD (like acute chest syndrome and splenic sequestration); however, a patient with SCD should also be evaluated for other causes of pain that are commonly seen in any patient who presents with these symptoms, like appendicitis. In the post-test a majority of subjects did choose the correct response (54%), however the remainder of post-test subjects (46%) thought the correct answer was answer 1, to monitor only for common SCD co-morbidites. This lowered the percentage of correct responses from 60% in the pre-test to 54% in the post-test. There are several possible explanations for this unexpected, post-test response. The storyline presented in the educational module, describes how the patient, Terrell, ended up with actue chest syndrome. Additionally there is a didactic segment reinforcing the importance of monitoring for co-morbid conditions like acute chest syndrome in patients with SCD. Furthermore, the physical exam that was performed on Terrell upon his acute abdominal pain symptoms may not have been sufficient to drive home the point that patients must still be monitored for any cause of his pain symptoms. This unexpected twist displayed in knowledge question 1 is an indication that time must be spent re-writing this portion of the case to clarify the need to monitor patients with SCD for other

causes for their symptoms that are unrelated to their disease. Lastly, fewer subjects actually took the post-test than the pre-test. It is possible that learners who accurately knew the pre-test answers chose not to take the time to take the post-test, shifting the post-test study population to a group with less experience in this scenario that may not pick up on the reasoning for the additional physical exam described in the case.

Knowledge question 2 and knowlegde question 3 showed similar findings. Question 2 asked what was the best way to determine initial dosing of pain medications for SCD patients. Question 3 asked what the goal of pain management was and what role opioids play in treating pain episodes in patients with SCD. The vast majority of the pre-test population correctly answered both questions, with slightly more answering correctly in the post-test. These questions show that there is high preliminary knowledge that using Indiviudalized Management Plans or institutional guidelines are the most effective methods for initial dosing, and that the overall goal of pain management is that aggressive treatment of pain episodes can help to reduce hospitalizations in this patient population.

Knowledge question 4 asked about the intervals for administration of pain medications, and yielded the most varied answer responses among the pre-test population. The most common answer in the pre-test was the correct response, that administration intervals follow assessment of patient's pain every 20 minutes. Post-test performance was improved, with 13% more subjects answering correctly. However, aside from knowledge question 1, question 4 had the highest post-test responses incorrect (31%). This is somewhat troubling as a great effort was made to reinforce constant reassessment of patient pain every 15-30 minutes to determine follow-up dosing. These persistant wrong answers could be a result of the wording of the question. Perhaps instead of "…at what intervals can pain medications be administered…" the question

should be more complete and read, "…at what intervals should a patient be reassessed to determine what follow-up doses should be administered…". Or, perhaps there should be more emphasis in the case that reassessment of the patients pain level every 15-30 minutes should involve administration of more pain medications if pain has not shown improvement. Changing wording within the case and for this knowledge question may drive home the point that management must involve patient input and should be aggressive.

Knowledge question 5 showed the biggest shift in correct responses between the pre-test (62%) and post-test (100%). This shift in knowledge could have been enhanced due to a similar question embedded within the case, that asked whether Terrell should be awoken to assess his pain, the answer was yes, he should. Another consideration for the nice improvement in correct responses could be a result of how the question was worded. Of all the knowledge questions it is the most succintly written, without room for misinterpretation.

Based on responses to evaluation question 5, a majority of subjects felt their level of knowledge of pain management was enhanced to some degree. However, answer selection from the pre and post test show that there remains room for improvement of knowledge. This will be achieved by careful re-wording of the knowledge questions, and deeper explanation of key learning points that were not adequately described or explicitly stated.

5.3 AIM 2: DETERMINE WHETHER HEALTH CARE PROVIDERS WILL ALTER THEY WAY THEY MANAGE PATIENTS WITH SCD PRESENTING TO THE ED OR TO THEIR CLINICAL PRACTICE WITH PAIN FOLLOWING THE USE OF THIS EDUCATIONAL MODULE

To elucidate whether subjects would change their approach or pratice of managing pain in patients with SCD examination of responses to evaluation question 6, “To what extent did the activity change the way you think about treating pain in patients with Sickle cell disease?”, evaluation 7, “To what extent will you make a change in your practice of treating pain in patients with Sickle cell disease as a result of your participation in this educational activity?”, and evaluation question 9, “Which of the following best describes the impact of this activity on how you will treat pain in patients with Sickle cell disease?” were considered.

As described above study subjects did indicate some enhanced kowledge. However, enhancement of knowledge did not cause a change in the way subjects thought about pain management. 50% of subjects felt they thought differently about pain management only to a “low” degree. Another 25% felt their change in practice would be to a “moderate” degree. The remaining 25% thought differently to a “high” or “very high” degree. An explanation for the many “low” or “moderate” answers is that this particular question could have been interpreted as whether the module altered attitudes toward treating pain managmement in patients with SCD, or altered future approach of treating pain. Evaluation question 6 would provide more information if it was re-worded to specifically ask about subjects attitudes about managing pain in patients with SCD.

Evaluation question 7 asks whether the subject will change the way they practice pain management. 92% said they would make a "low" or "moderate" change in practice. Given the moderate shifts in knowledge and mind-set of pain management, it is not surprising that subjects indicate minor changes in their practice of pain management as well. Evaluation question 9 asks subjects to describe how and why practice will or will not change following this educational activity. 73% (8/11 responses) said they will not change because they are already practicing as the educational module recommends. While this is reassuring information, it could indicate sample bias amoung our subjects. Our recruitment methods included dissemination of this module to various groups and institutions who tend to be well versed in the most up-to-date treatment practices for SCD and the best pain management recommendations. Indeed, the treatment protocol described in our educational module, and the algorithm created by the CHP Comprehensive Hemoglobinopathy Program, is compiled by the latest and most recent recommendations by experts in the field of SCD.

5.4 AIM 3: TO DETERMINE HEALTH CARE PROVIDERS OVERALL ATTITUDES ABOUT WEB-BASED EDUCATIONAL MODULES

A majority (41%) of subjects were satisified with the overall quality of the educational module to a "high" degree, followed by 35% who were satisfied to a "very high" degree. 24% of subjects were satisfied with the quality to a moderate degree. Similar responses were given when asked about the relevance of the content of the educational module to subjects' practice, with 81% of subjects feeling content was relevant to a "very high" or "high" degree. Additionally, 80% of

subjects felt that this type of web-based education was useful to a “very high” or “high” degree. Of note, more subjects indicated that the felt web-based modules were useful than there were subjects who felt they had gained knowledge by using this module. This information is significant; despite variable degrees of knowledge enhancement, subjects indicated that this type of educational tool is well received by learners. This is promising that as content refinement continues, learners will also continue to take advantage of this educational application.

5.5 AIM 4: ASSESS WHETHER THIS TYPE OF EDUCATIONAL TOOL HELPED PROVIDERS TO LEARN AND UNDERSTAND THE ESTABLISHED CHP ALGORITHM FOR MANAGING VASOOLUSIVE PAIN IN PATIENTS WITH SICKLE CELL DISEASE

64% of subjects reported a “moderate” shift in learning and understanding the CHP algorithm for managing pain in SCD. The remaining 36% felt the module help to understand the algorithm to a “very high” or “high” degree. When asked whether the educational objectives were achieved in the module, 36% subjects reported the objectives were at least “moderately” achieved, the remaining subjects felt the objective were achieved to a “high” or “very high” degree. We were expecting similar responses to these two evaluation questions as the educational objectives were based on the algorithm. The difference between responses to these questions may be due to the fact that the learning objectives were clearly listed on one of the first slides of the module. The algorithm is given in the form of a PDF link and users are asked to refer to it while going through the case. It is possible, although unlikely, that some users do not have software like, Adobe on

their computers and were therefore unable to access the algorithm. More likely is the possibility that the algorithm is not being printed by all subjects and is instead glanced over briefly by the learner. Due to the degree of information in the algorithm, a one-time glance may not be enough to grasp all the content included in this algorithm. Manipulations to the module in the future could include a way to provide access to the algorithm periodically throughout the module. Additionally, the number of "moderate" responses indicates that there are topics that must be expanded upon in order to clarify the algorithm and learning objectives.

5.6 LIMITATIONS AND OTHER CONSIDERATIONS

The announcement of the availability of this educational module for health-care providers (potential study subjects) began on March 8, 2007, leaving about three weeks for data collection. As a result there is a very small sample size with number of subjects varying with each question, ranging from 11-20 responses. The limited number of responses makes it difficult to interpret the data collected thus far. The case will remain accessible via CHP's Sickle Cell Program web page and on the PedsEd site at least for the next year. Furthermore, the case can be located on Google as the second, third, and fourth response by searching, "sickle cell Pittsburgh". It is our hope that this wide accessibility to the case will allow for continual data collection.

In order to maintain IRB requirements, including anonymity and option to not participate, users were able to skip any questions they wanted. Due to the nature of a web-based application, and the fact that our subjects are health-care providers, it is not difficult to imagine how often someone may be interrupted, and for whatever reason fail to answer all the questions asked. In

an effort to maintain anonymity and provide easy, quick access to the module, learners were not required to log-in to the educational module or to PedsEd to access the case.

Not requiring learners to log-in to the educational module does provide anonymity and easy access for learners. However, this feature also prevents the ability to follow one learner's responses through the entire module; from pre-test, to post-test, through the evaluation. The programming necessary to create the custom reports that could follow individual users, simply would take too much time to create, or would inevitably require users to log-in with a disclaimer that their information would be de-identified prior to data collection. In either case, we would loose some of our study population.

The inability to track one learners answer responses through the module is limiting for two reasons. First, it prevents the ability to make any conclusions about whether certain answer choices track with demographic information, or work experience. Second, we are unable to see how individuals improved with the knowledge questions. Clearly there are more answer responses for the pre-test, than for the post-test. If we had been able to track individuals we would also be able to see if there was any correlation between pre-test scores and not completing the remainder of the questions. Without being able to track demographic information and pattern of responses or measure individual learner's pre and post test scores, we were unable to perform any statistical analysis.

As previously mentioned, in an effort to disperse this educational module to as many clinicians as possible, the range of background knowledge varies a great deal. By dispersing this educational tool to many health care providers, we are able to receive feedback from individuals with various perspectives, which is essential. On the other hand it becomes difficult to interpret data, especially in light of our small sample size. For example, were the four individuals who

continued to get knowledge question 4 wrong in the post-test answered by individuals who received their medical or nursing degree last year, or were they answered by Hematology attendings. The answer has very different implications.

Much of our recruitment information went out to health care providers who are experts in the field. Although this ensures useful and constructive feedback, it does add the potential sample bias by a population of subjects that are SCD experts. Looking forward, as our subject population increases it would be of interest to determine how comfortable health care providers feel treating pain in SCD and administering opioids prior to participating in this educational module and then ask a similar question at the completion of the module. This would determine whether confidence level in treating patients with opioids and relates to level of knowledge.

5.7 FUTURE STUDIES

Moving forward, we will continue to refine case content, as well as in the way the knowledge and evaluation questions are written. Content of the case will be improved to increase delineation of teaching points, and to reinforce learning objectives. Both knowledge and evaluation questions will be edited to allow for more succinct, concise, and relevant questions.

Other areas that could be addressed involve the technology side of the educational module. We will have to determine a method to track each learner's responses without using their personal information and maintaining ease of use of the module. Perhaps each user could be assigned an ID as soon as they answer the first knowledge pre-test question, or instead

learners could be asked to give themselves any name or ID on the same screen where the learner is told to imagine they are a pediatric Emergency Medicine resident.

To increase awareness and understanding of Children's Hospital of Pittsburgh's Guidelines for Clinical Effectiveness for managing pain from vaso-occlusion in patient's with Sickle cell disease, we will provide continued access to the algorithm throughout the case. This will provide convenient availability of the algorithm as learners answer the question sets.

Currently, learners are given the option to not respond to some or any questions asked in order to comply with IRB research requirements. While we continue to collect data, consent for participation by the learner will be elicited prior to beginning the module. Once an learner has agreed to participate they will be required to answer all questions. In addition, the post-test questions may be moved from their current location at the very end of the case to interspersing them throughout the case, much the informational, embedded questions. By inserting the post-test questions within the case where the correct answer is being discussed will put the question in context, and provide reinforcement of that particular teaching point.

In the future, it is possible that this module or modules similar to this one could be incorporated into medical school curriculums, or available for continuing education credits. In either of these scenarios, users will be required to answer all questions within the module. This paired with the ability to track one subject's answer responses will allow for more complete and accurate data analysis.

6.0 CONCLUSIONS

Subjects achieved moderate enhancement of knowledge following the use of this educational module. Due to the limitations in this study it is difficult to distinguish if this is because learners had a high level of preliminary knowledge, if the pre and post test subject populations varied significantly in their background knowledge, or if it is tied to the wording of case content or the questions asks. To eliminate one of these possibilities efforts will involve revisions in the case to clarify and emphasize teaching points, and re-wording to streamline questions.

The majority of subjects indicated that they would make few changes in the way they actively manage pain in patients with SCD. This coincides with moderate gains in knowledge and in understanding of the CHP algorithm (Guidelines for Clinical Effectiveness) for managing vaso-occlusive pain in SCD patients. Interestingly, most learners feel they are already treating patients in the manner described by our project. Again, this indicates efforts must be spent reinforcing the primary learning objectives of our educational module. Subjects did indicate that they felt that the module was of good quality, relevant to their work, and felt is was a useful mode of education.

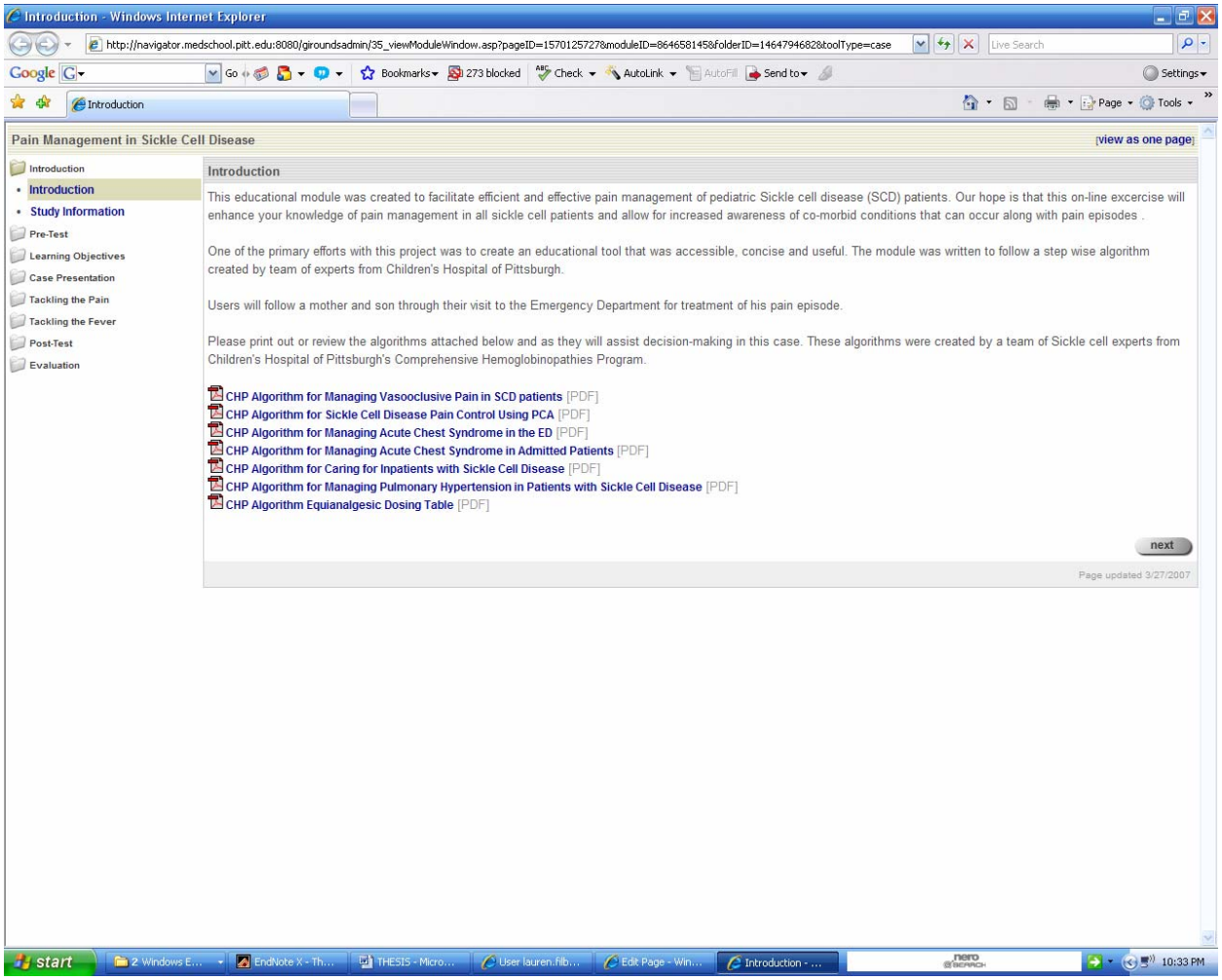
Based on the information provided by this project, an interactive web-based educational module can enhance health-care providers' knowledge of pain management in SCD with additional efforts to improve content and question sets. A web-based application is appealing to

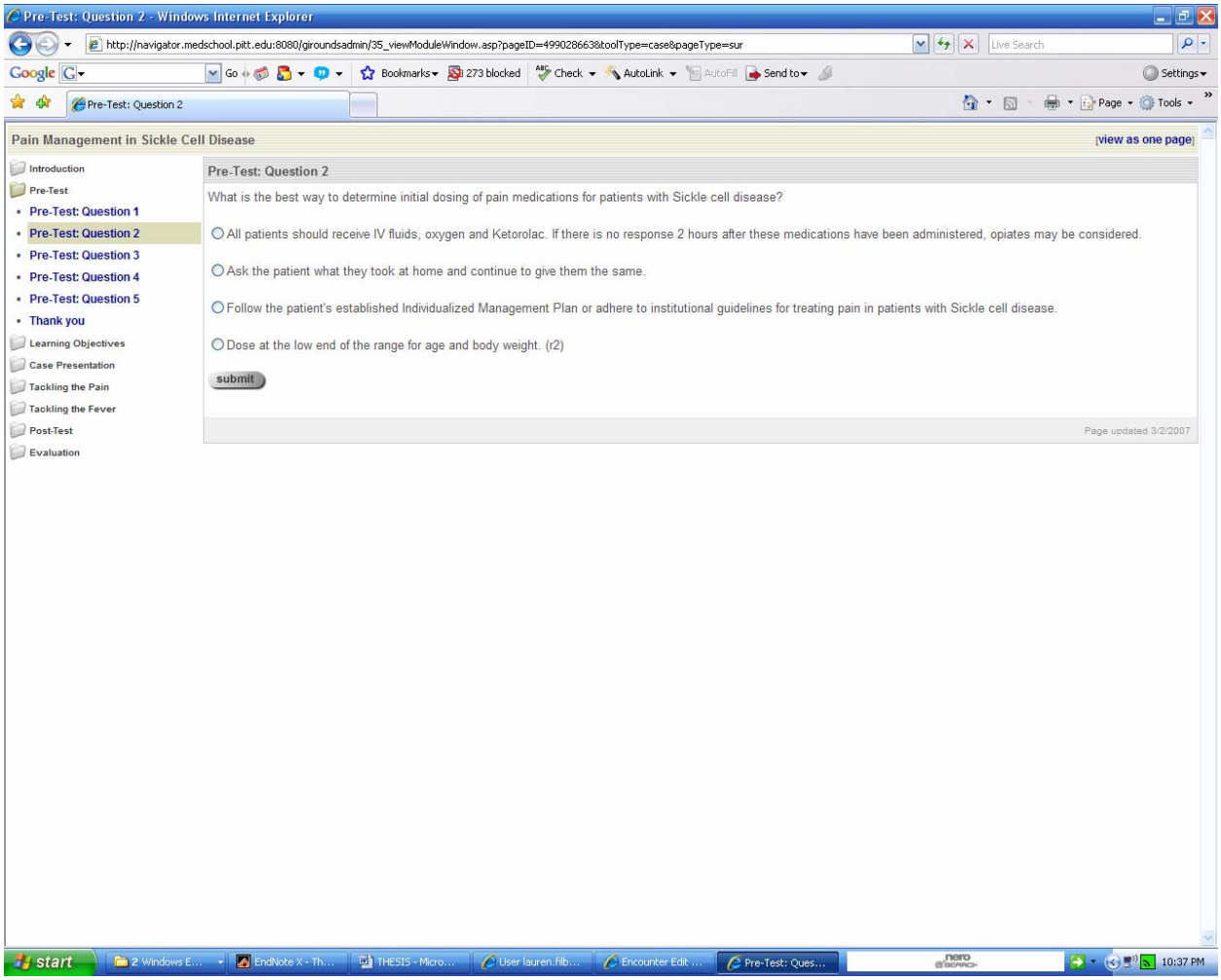
learners, and can aid in providers becoming more familiar with the institutional Guidelines for Clinical Effectiveness.

From a public health perspective, undertreatment of pain in health care settings is a major problem. In SCD, undertreatment of pain episodes causes increased hospital admission rates and decreased overall quality of life. Poorly managed pain in SCD can affect both physical and emotional well-being. Enhancing health-care providers' knowledge and awareness of effective methods of managing pain in SCD is the key to beginning to alleviate this problem. As health care providers improve their practices, patient anxiety will reduce, hospitalizations will decrease, and inevitably, quality of life will improve for many patients.

APPENDIX A

SELECTED SCREEN SHOTS OF EDUCATIONAL MODULE

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Pre-Test: Question 2
Pain Management in Sickle Cell Disease
[view as one page]
Introduction
Pre-Test
Pre-Test: Question 1
Pre-Test: Question 2
Pre-Test: Question 3
Pre-Test: Question 4
Pre-Test: Question 5
Thank you
Learning Objectives
Case Presentation
Tackling the Pain
Tackling the Fever
Post-Test
Evaluation
Pre-Test: Question 2
What is the best way to determine initial dosing of pain medications for patients with Sickle cell disease?
All patients should receive IV fluids, oxygen and Ketorolac. If there is no response 2 hours after these medications have been administered, opiates may be considered.
Ask the patient what they took at home and continue to give them the same.
Follow the patient's established Individualized Management Plan or adhere to institutional guidelines for treating pain in patients with Sickle cell disease.
Dose at the low end of the range for age and body weight. (r2)
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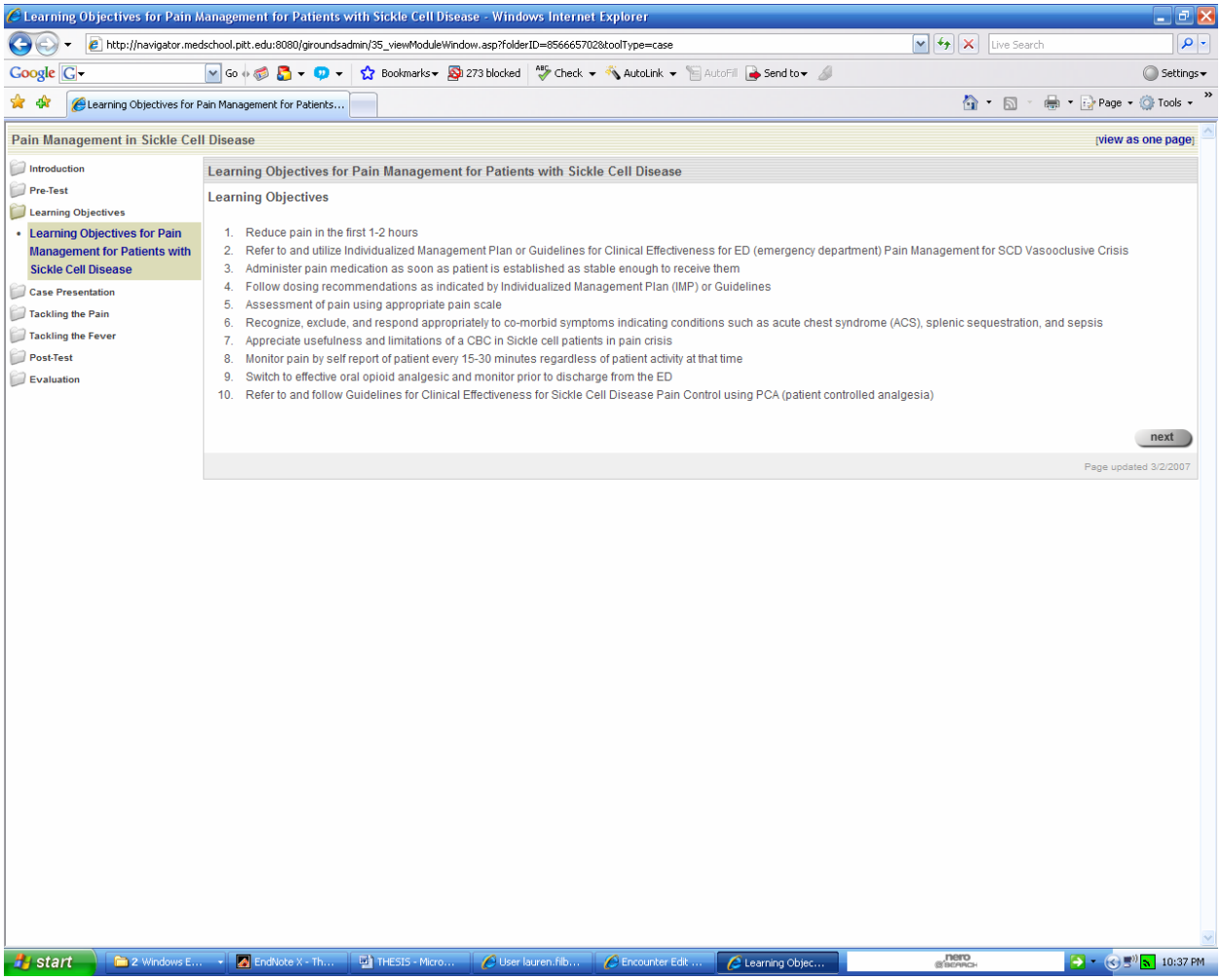
Learning Objectives for Pain Management for Patients with Sickle Cell Disease - Windows Internet Explorer
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Pain Management in Sickle Cell Disease
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Learning Objectives for Pain Management for Patients with Sickle Cell Disease
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Tackling the Pain
Tackling the Fever
Post-Test
Evaluation
Learning Objectives for Pain Management for Patients with Sickle Cell Disease
Learning Objectives
1. Reduce pain in the first 1-2 hours
2. Refer to and utilize Individualized Management Plan or Guidelines for Clinical Effectiveness for ED (emergency department) Pain Management for SCD Vasoocclusive Crisis
3. Administer pain medication as soon as patient is established as stable enough to receive them
4. Follow dosing recommendations as indicated by Individualized Management Plan (IMP) or Guidelines
5. Assessment of pain using appropriate pain scale
6. Recognize, exclude, and respond appropriately to co-morbid symptoms indicating conditions such as acute chest syndrome (ACS), splenic sequestration, and sepsis
7. Appreciate usefulness and limitations of a CBC in Sickle cell patients in pain crisis
8. Monitor pain by self report of patient every 15-30 minutes regardless of patient activity at that time
9. Switch to effective oral opioid analgesic and monitor prior to discharge from the ED
10. Refer to and follow Guidelines for Clinical Effectiveness for Sickle Cell Disease Pain Control using PCA (patient controlled analgesia)
next
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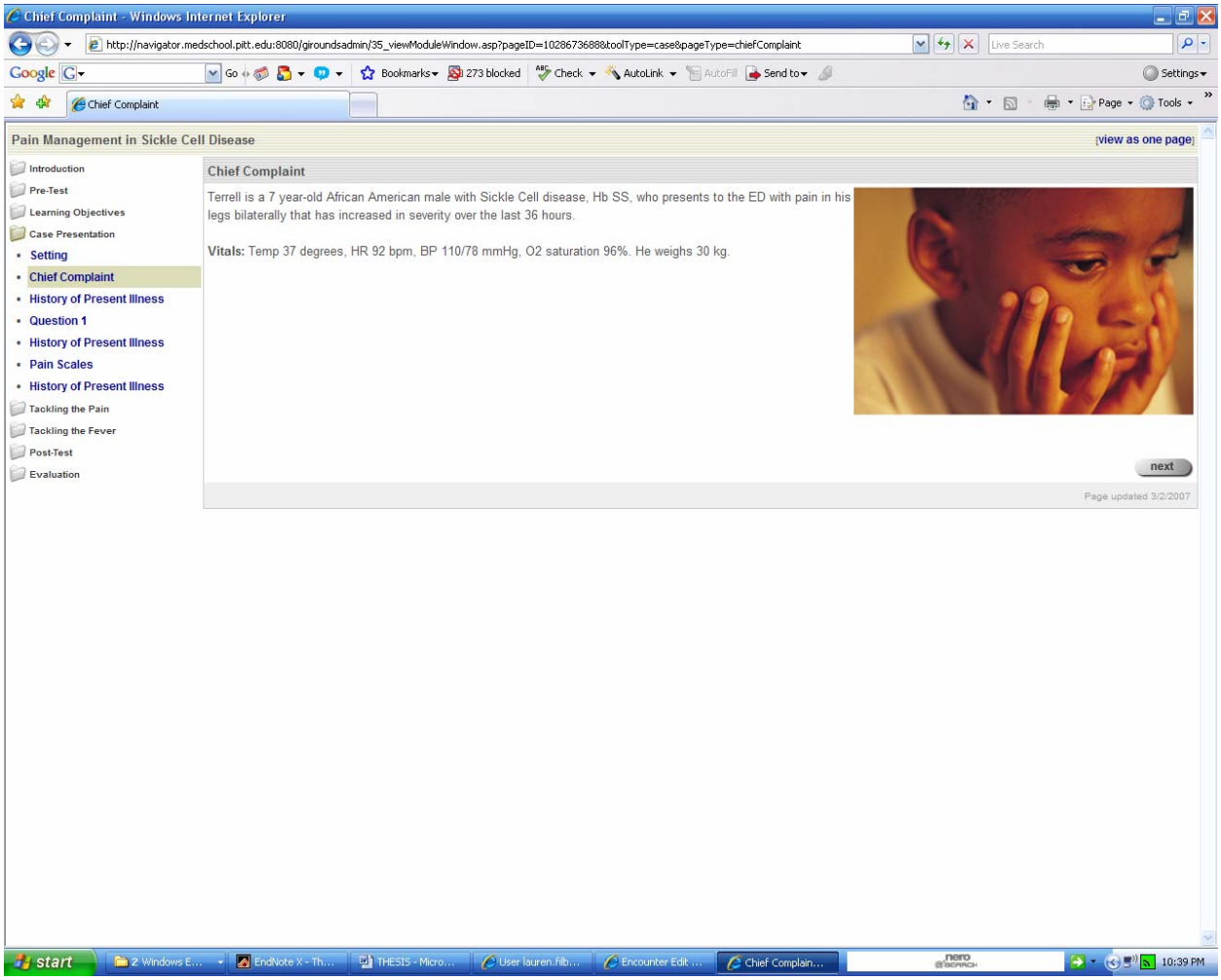
Chief Complaint - Windows Internet Explorer
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Pain Management in Sickle Cell Disease
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Setting
Chief Complaint
History of Present Illness
Question 1
History of Present Illness
Pain Scales
History of Present Illness
Tackling the Pain
Tackling the Fever
Post-Test
Evaluation
Chief Complaint
Terrell is a 7 year-old African American male with Sickle Cell disease, Hb SS, who presents to the ED with pain in his legs bilaterally that has increased in severity over the last 36 hours.
Vitals: Temp 37 degrees, HR 92 bpm, BP 110/78 mmHg, O2 saturation 96%. He weighs 30 kg.
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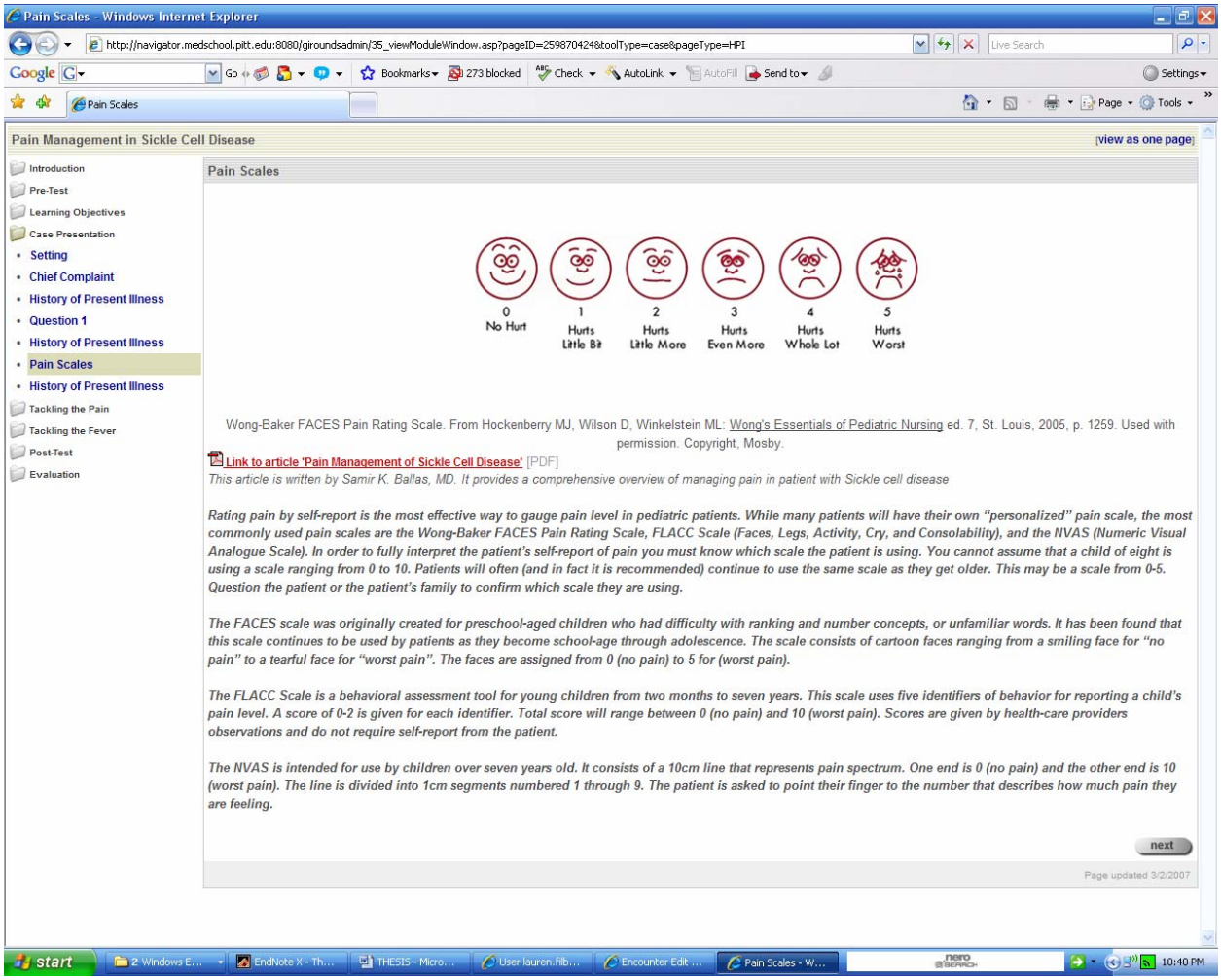
Pain Scales - Windows Internet Explorer
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History of Present Illness
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Tackling the Fever
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Evaluation
Pain Scales
0 No Hurt
1 Hurts Little Bit
2 Hurts Little More
3 Hurts Even More
4 Hurts Whole Lot
5 Hurts Worst
Wong-Baker FACES Pain Rating Scale. From Hockenberry MJ, Wilson D, Winkelstein ML: Wong's Essentials of Pediatric Nursing ed. 7, St. Louis, 2005, p. 1259. Used with permission. Copyright, Mosby.
Link to article 'Pain Management of Sickle Cell Disease' [PDF]
This article is written by Samir K. Ballas, MD. It provides a comprehensive overview of managing pain in patient with Sickle cell disease
Rating pain by self-report is the most effective way to gauge pain level in pediatric patients. While many patients will have their own "personalized" pain scale, the most commonly used pain scales are the Wong-Baker FACES Pain Rating Scale, FLACC Scale (Faces, Legs, Activity, Cry, and Consolability), and the NVAS (Numeric Visual Analogue Scale). In order to fully interpret the patient's self-report of pain you must know which scale the patient is using. You cannot assume that a child of eight is using a scale ranging from 0 to 10. Patients will often (and in fact it is recommended) continue to use the same scale as they get older. This may be a scale from 0-5. Question the patient or the patient's family to confirm which scale they are using.
The FACES scale was originally created for preschool-aged children who had difficulty with ranking and number concepts, or unfamiliar words. It has been found that this scale continues to be used by patients as they become school-age through adolescence. The scale consists of cartoon faces ranging from a smiling face for "no pain" to a tearful face for "worst pain". The faces are assigned from 0 (no pain) to 5 for (worst pain).
The FLACC Scale is a behavioral assessment tool for young children from two months to seven years. This scale uses five identifiers of behavior for reporting a child's pain level. A score of 0-2 is given for each identifier. Total score will range between 0 (no pain) and 10 (worst pain). Scores are given by health-care providers observations and do not require self-report from the patient.
The NVAS is intended for use by children over seven years old. It consists of a 10cm line that represents pain spectrum. One end is 0 (no pain) and the other end is 10 (worst pain). The line is divided into 1cm segments numbered 1 through 9. The patient is asked to point their finger to the number that describes how much pain they are feeling.
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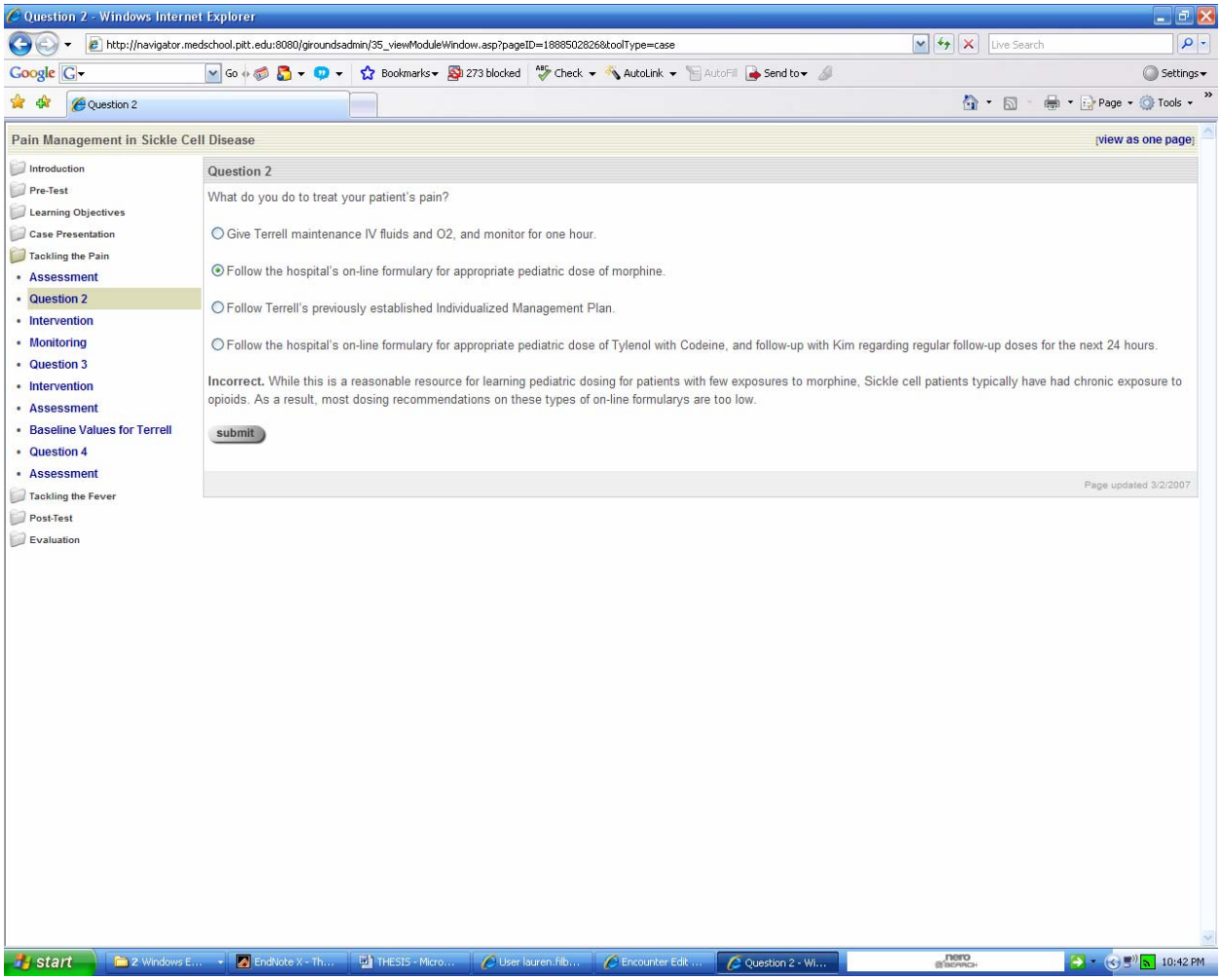
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Question 2
Pain Management in Sickle Cell Disease
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Question 3
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Assessment
Baseline Values for Terrell
Question 4
Assessment
Tackling the Fever
Post-Test
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Question 2
What do you do to treat your patient's pain?
Give Terrell maintenance IV fluids and O2, and monitor for one hour.
Follow the hospital's on-line formulary for appropriate pediatric dose of morphine.
Follow Terrell's previously established Individualized Management Plan.
Follow the hospital's on-line formulary for appropriate pediatric dose of Tylenol with Codeine, and follow-up with Kim regarding regular follow-up doses for the next 24 hours.
Incorrect. While this is a reasonable resource for learning pediatric dosing for patients with few exposures to morphine, Sickle cell patients typically have had chronic exposure to opioids. As a result, most dosing recommendations on these types of on-line formularys are too low.
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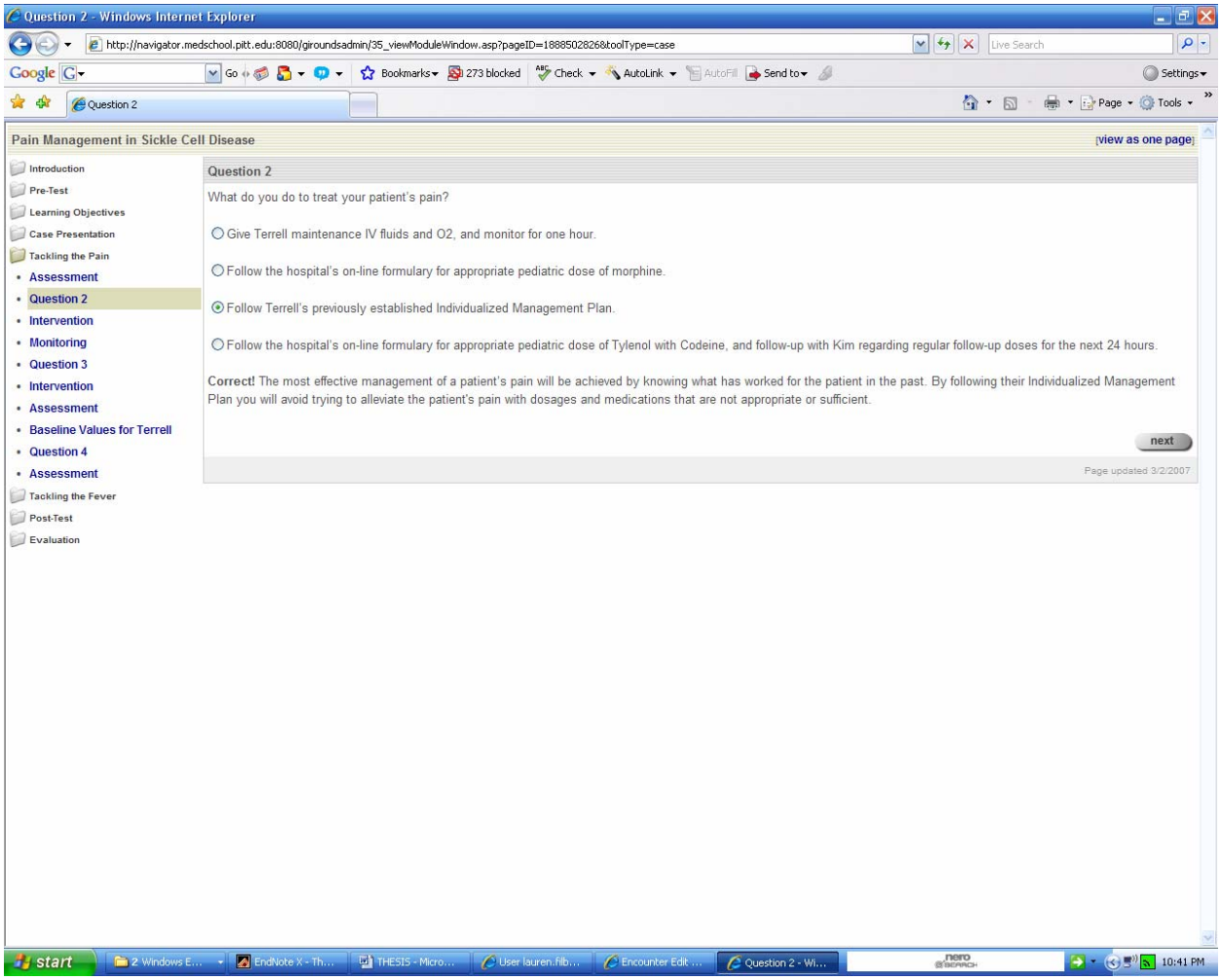
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Follow the hospital's on-line formulary for appropriate pediatric dose of morphine.
Follow Terrell's previously established Individualized Management Plan.
Follow the hospital's on-line formulary for appropriate pediatric dose of Tylenol with Codeine, and follow-up with Kim regarding regular follow-up doses for the next 24 hours.
Correct! The most effective management of a patient's pain will be achieved by knowing what has worked for the patient in the past. By following their Individualized Management Plan you will avoid trying to alleviate the patient's pain with dosages and medications that are not appropriate or sufficient.
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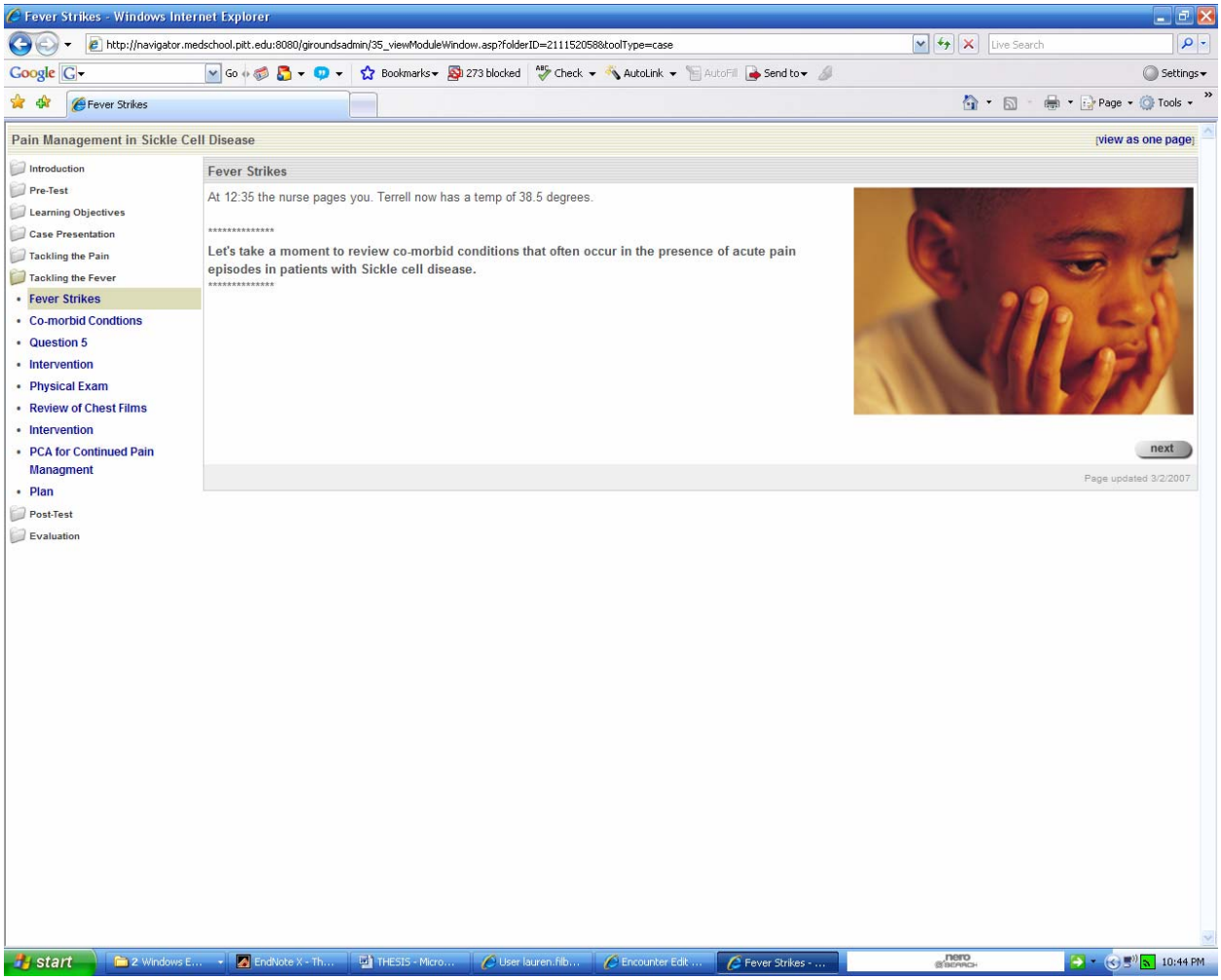
Fever Strikes - Windows Internet Explorer
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Fever Strikes
At 12:35 the nurse pages you. Terrell now has a temp of 38.5 degrees.
****
Let's take a moment to review co-morbid conditions that often occur in the presence of acute pain episodes in patients with Sickle cell disease.
****
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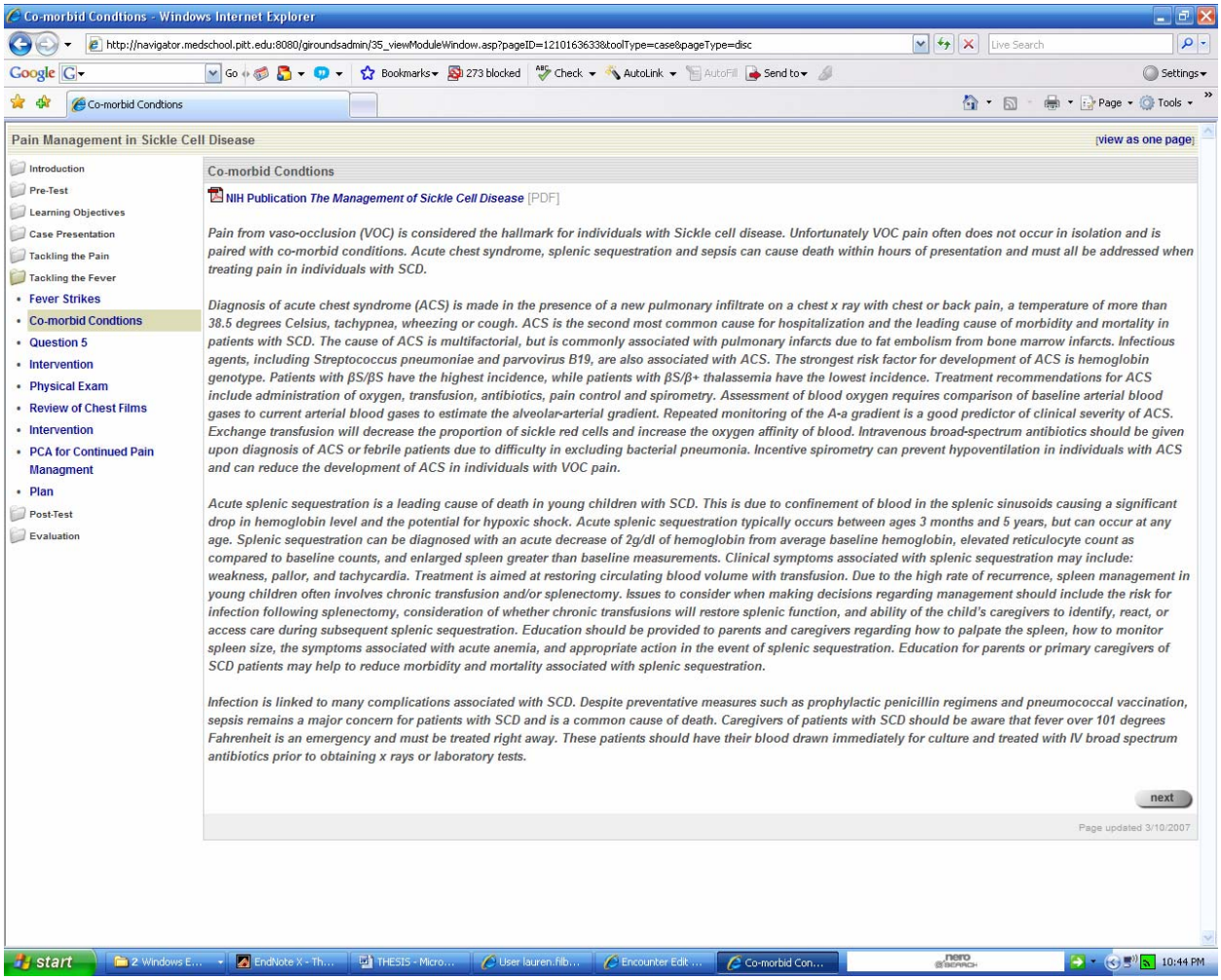
Co-morbid Condtions - Windows Internet Explorer
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NIH Publication The Management of Sickle Cell Disease [PDF]
Pain from vaso-occlusion (VOC) is considered the hallmark for individuals with Sickle cell disease. Unfortunately VOC pain often does not occur in isolation and is paired with co-morbid conditions. Acute chest syndrome, splenic sequestration and sepsis can cause death within hours of presentation and must all be addressed when treating pain in individuals with SCD.
Diagnosis of acute chest syndrome (ACS) is made in the presence of a new pulmonary infiltrate on a chest x ray with chest or back pain, a temperature of more than 38.5 degrees Celsius, tachypnea, wheezing or cough. ACS is the second most common cause for hospitalization and the leading cause of morbidity and mortality in patients with SCD. The cause of ACS is multifactorial, but is commonly associated with pulmonary infarcts due to fat embolism from bone marrow infarcts. Infectious agents, including Streptococcus pneumoniae and parvovirus B19, are also associated with ACS. The strongest risk factor for development of ACS is hemoglobin genotype. Patients with βS/βS have the highest incidence, while patients with βS/β+ thalassemia have the lowest incidence. Treatment recommendations for ACS include administration of oxygen, transfusion, antibiotics, pain control and spirometry. Assessment of blood oxygen requires comparison of baseline arterial blood gases to current arterial blood gases to estimate the alveolar-arterial gradient. Repeated monitoring of the A-a gradient is a good predictor of clinical severity of ACS. Exchange transfusion will decrease the proportion of sickle red cells and increase the oxygen affinity of blood. Intravenous broad-spectrum antibiotics should be given upon diagnosis of ACS or febrile patients due to difficulty in excluding bacterial pneumonia. Incentive spirometry can prevent hypoventilation in individuals with ACS and can reduce the development of ACS in individuals with VOC pain.
Acute splenic sequestration is a leading cause of death in young children with SCD. This is due to confinement of blood in the splenic sinusoids causing a significant drop in hemoglobin level and the potential for hypoxic shock. Acute splenic sequestration typically occurs between ages 3 months and 5 years, but can occur at any age. Splenic sequestration can be diagnosed with an acute decrease of 2g/dl of hemoglobin from average baseline hemoglobin, elevated reticulocyte count as compared to baseline counts, and enlarged spleen greater than baseline measurements. Clinical symptoms associated with splenic sequestration may include: weakness, pallor, and tachycardia. Treatment is aimed at restoring circulating blood volume with transfusion. Due to the high rate of recurrence, spleen management in young children often involves chronic transfusion and/or splenectomy. Issues to consider when making decisions regarding management should include the risk for infection following splenectomy, consideration of whether chronic transfusions will restore splenic function, and ability of the child's caregivers to identify, react, or access care during subsequent splenic sequestration. Education should be provided to parents and caregivers regarding how to palpate the spleen, how to monitor spleen size, the symptoms associated with acute anemia, and appropriate action in the event of splenic sequestration. Education for parents or primary caregivers of SCD patients may help to reduce morbidity and mortality associated with splenic sequestration.
Infection is linked to many complications associated with SCD. Despite preventative measures such as prophylactic penicillin regimens and pneumococcal vaccination, sepsis remains a major concern for patients with SCD and is a common cause of death. Caregivers of patients with SCD should be aware that fever over 101 degrees Fahrenheit is an emergency and must be treated right away. These patients should have their blood drawn immediately for culture and treated with IV broad spectrum antibiotics prior to obtaining x rays or laboratory tests.
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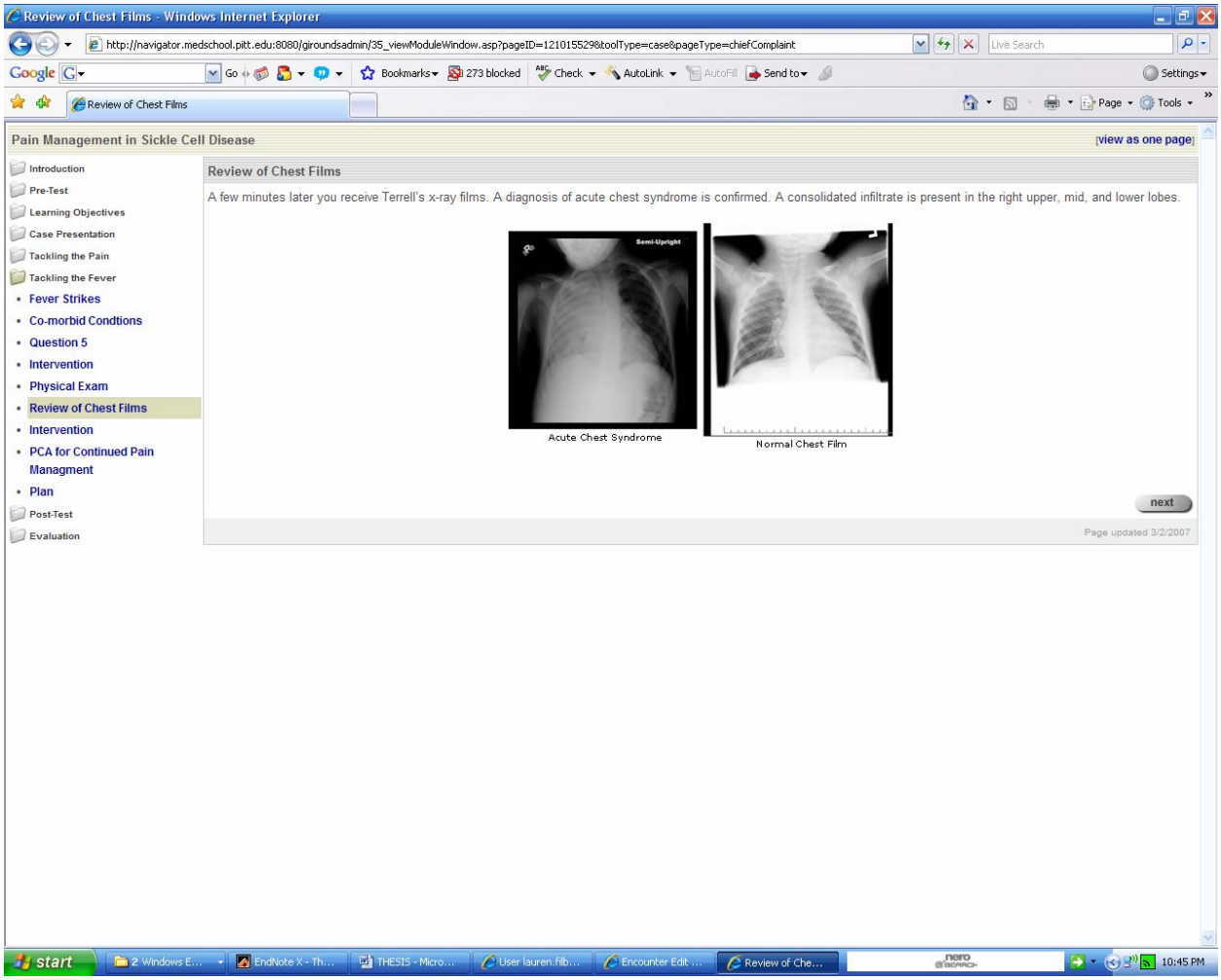
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A few minutes later you receive Terrell's x-ray films. A diagnosis of acute chest syndrome is confirmed. A consolidated infiltrate is present in the right upper, mid, and lower lobes.
Semi-Upright
Acute Chest Syndrome
Normal Chest Film
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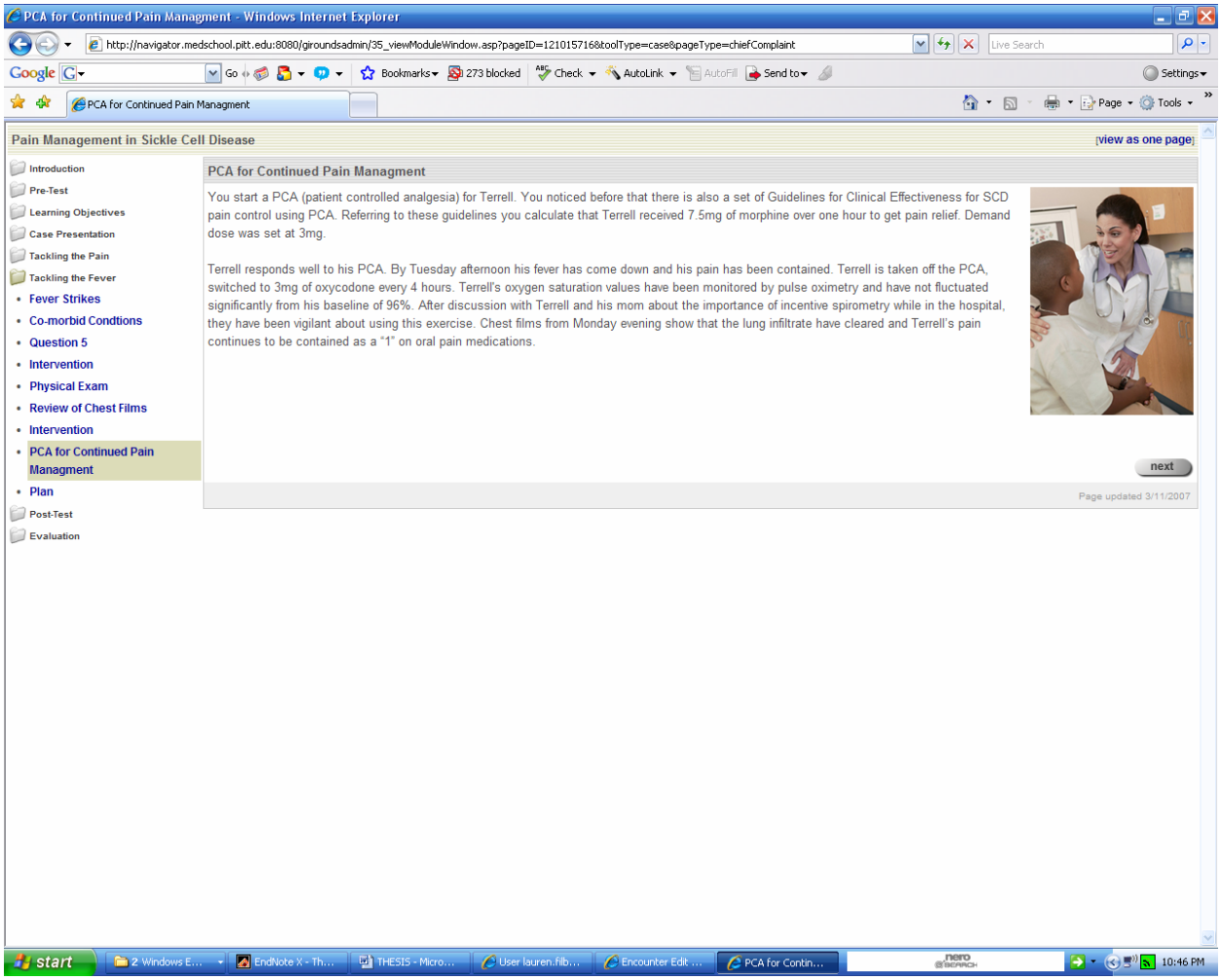
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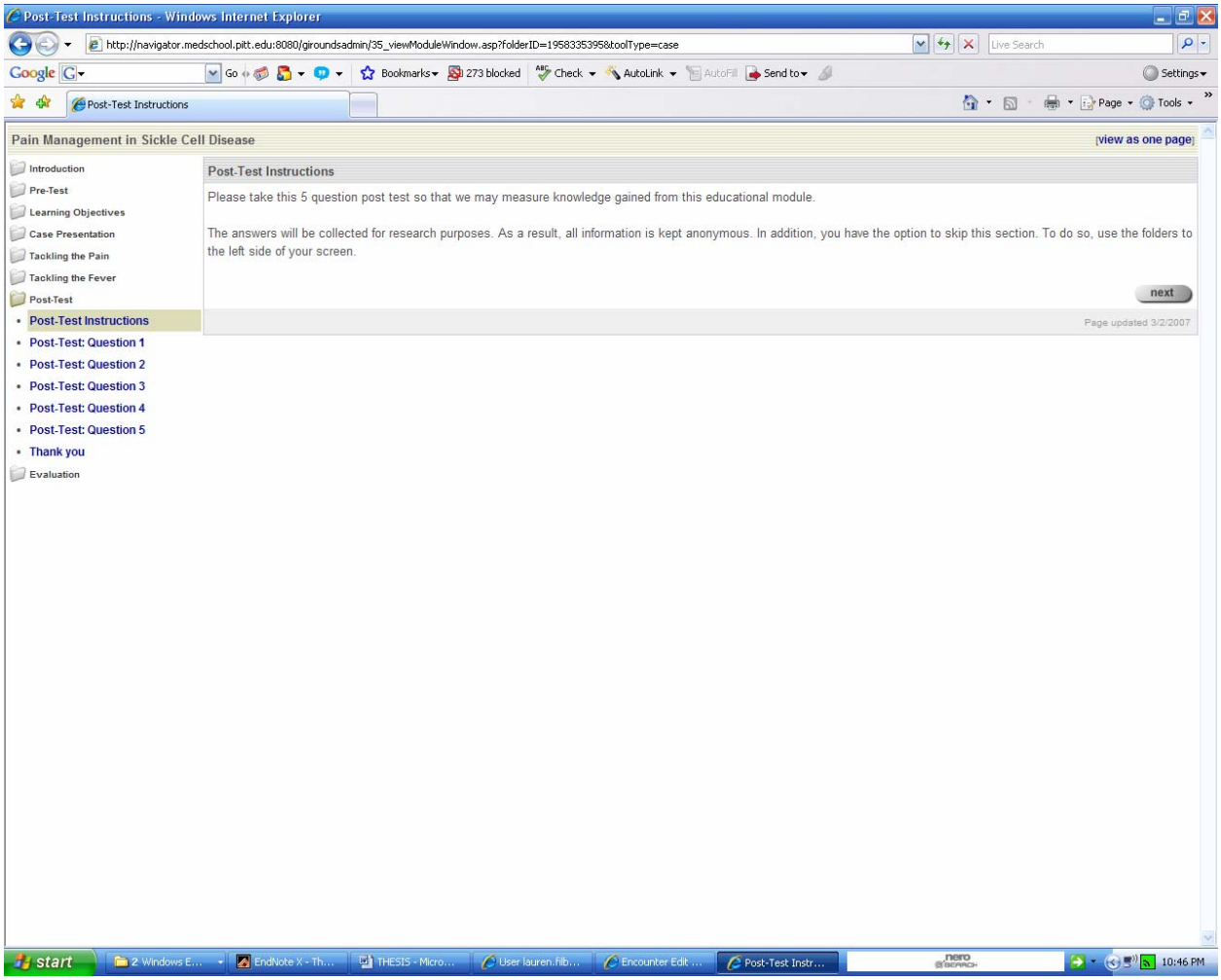
Evaluation
PCA for Continued Pain Managment
You start a PCA (patient controlled analgesia) for Terrell. You noticed before that there is also a set of Guidelines for Clinical Effectiveness for SCD pain control using PCA. Referring to these guidelines you calculate that Terrell received 7.5mg of morphine over one hour to get pain relief. Demand dose was set at 3mg.
Terrell responds well to his PCA. By Tuesday afternoon his fever has come down and his pain has been contained. Terrell is taken off the PCA, switched to 3mg of oxycodone every 4 hours. Terrell's oxygen saturation values have been monitored by pulse oximetry and have not fluctuated significantly from his baseline of 96%. After discussion with Terrell and his mom about the importance of incentive spirometry while in the hospital, they have been vigilant about using this exercise. Chest films from Monday evening show that the lung infiltrate have cleared and Terrell's pain continues to be contained as a "1" on oral pain medications.
next
Page updated 3/11/2007
start
10:46 PM


Post-Test Instructions - Windows Internet Explorer
http://navigator.medschool.pitt.edu:8080/giroundsadmin/35_viewModuleWindow.asp?folderID=1958335395&toolType=case
Pain Management in Sickle Cell Disease
[view as one page]
Introduction
Pre-Test
Learning Objectives
Case Presentation
Tackling the Pain
Tackling the Fever
Post-Test
Post-Test Instructions
Post-Test: Question 1
Post-Test: Question 2
Post-Test: Question 3
Post-Test: Question 4
Post-Test: Question 5
Thank you
Evaluation
Post-Test Instructions
Please take this 5 question post test so that we may measure knowledge gained from this educational module.
The answers will be collected for research purposes. As a result, all information is kept anonymous. In addition, you have the option to skip this section. To do so, use the folders to the left side of your screen.
next
Page updated 3/2/2007
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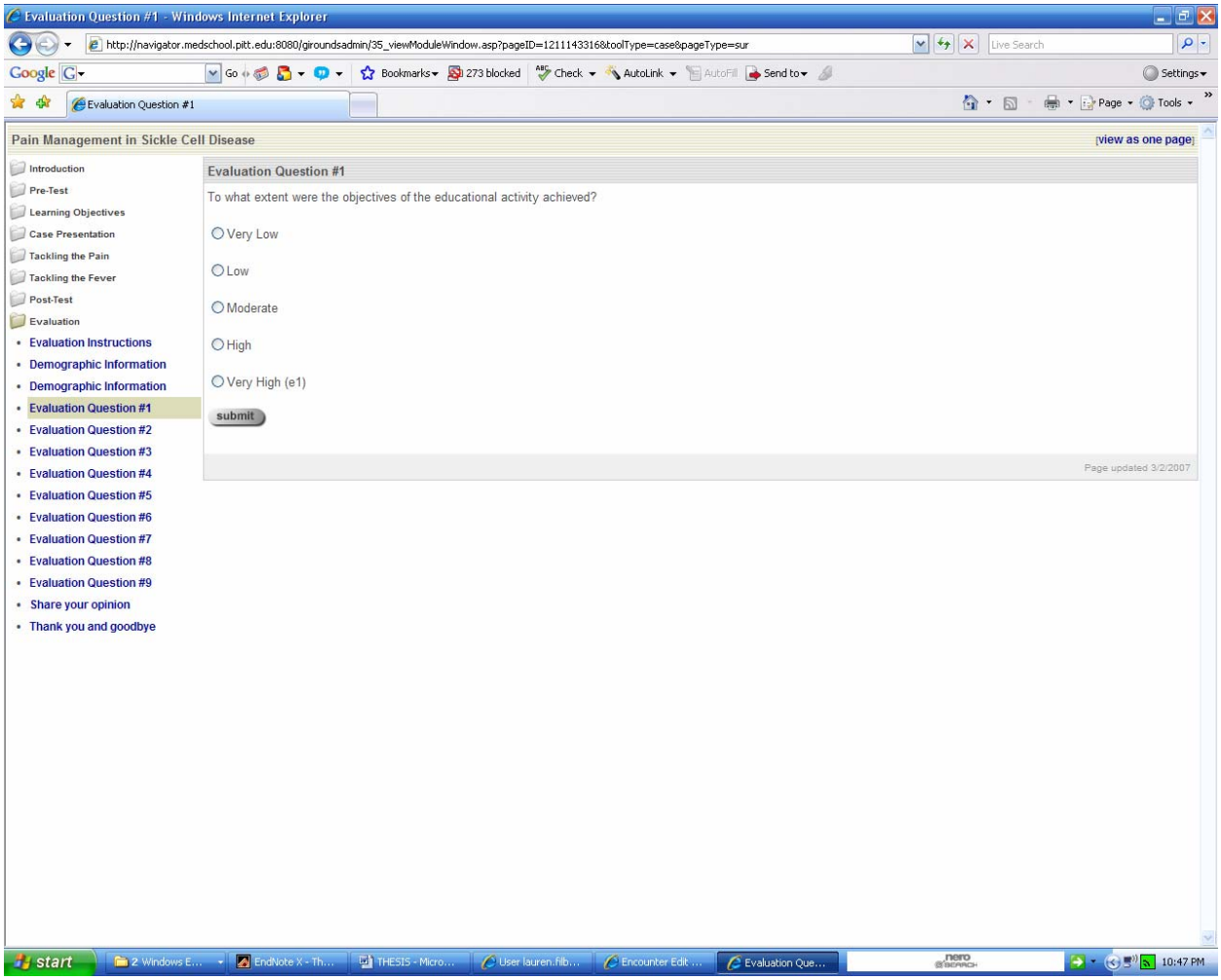
Evaluation Question #1 - Windows Internet Explorer
http://navigator.medschool.pitt.edu:8080/giroundsadmin/35_viewModuleWindow.asp?pageID=1211143316&toolType=case&pageType=sur
Pain Management in Sickle Cell Disease
[view as one page]
Introduction
Pre-Test
Learning Objectives
Case Presentation
Tackling the Pain
Tackling the Fever
Post-Test
Evaluation
Evaluation Instructions
Demographic Information
Demographic Information
Evaluation Question #1
Evaluation Question #2
Evaluation Question #3
Evaluation Question #4
Evaluation Question #5
Evaluation Question #6
Evaluation Question #7
Evaluation Question #8
Evaluation Question #9
Share your opinion
Thank you and goodbye
Evaluation Question #1
To what extent were the objectives of the educational activity achieved?
Very Low
Low
Moderate
High
Very High (e1)
submit
Page updated 3/2/2007
start
10:47 PM

APPENDIX B

IRB APPROVAL LETTER

University of Pittsburgh
Institutional Review Board

Exempt and Expedited Reviews

University of Pittsburgh FWA: 00006790
University of Pittsburgh Medical Center: FWA 00006735
Children's Hospital of Pittsburgh: FWA 00000600

3500 Fifth Avenue
Suite 100
Pittsburgh, PA 15213
Phone: 412.383.1480
Fax: 412.383.1508

TO: Lakshmanan Krishnamurti, M.D.

FROM: Christopher M. Ryan, PhD, Vice Chair

DATE: February 23, 2007

PROTOCOL: Evaluation of an Interactive Educational Module for Health Care Providers Treating Pain in Patients with Sickle Cell Disease

IRB Number: 0702021

The above-referenced protocol has been reviewed by the University of Pittsburgh Institutional Review Board. Based on the information provided in the IRB protocol, this project meets all the necessary criteria for an exemption, and is hereby designated as "exempt" under section 45 CFR 46.101(b)(2).

- If any modifications are made to this project, please submit an 'exempt modification' form to the IRB.
- Please advise the IRB when your project has been completed so that it may be officially terminated in the IRB database.
- This research study may be audited by the University of Pittsburgh Research Conduct and Compliance Office.

Approval Date: February 23, 2007

CR:kh

APPENDIX C

RECRUITMENT E-MAIL

Dear Colleague,

In an effort to facilitate efficient and effective pain management in patients with Sickle cell disease, we have created a comprehensive, web-based educational module for use by all health care providers who treat pain in patients with this complex disease.

The module provides an instantaneous link to peer reviewed information to effectively treat pain episodes in patients with Sickle cell disease. Users will follow a virtual clinic experience of a child with Sickle cell disease presenting to the Emergency Department with pain, and will learn to successfully manage his current pain episode. Users are asked to complete a brief test of knowledge of the key principles of vaso-occlusive pain management for a patient with sickle cell disease presenting to the Emergency Department. Once completing the module the user will be requested to complete it again. We also request that they complete a brief evaluation and invite additional feedback to determine overall opinion about this type of educational tool.

We are asking that you encourage your colleagues and other health care providers (including attending physicians, fellows, residents, nurses, psychologists, social workers, etc.) to take advantage of this unique educational opportunity.

Information gathered in the pre and post test and evaluation tool are collected anonymously and will be used for refinement and further improvement of this educational module.

Please visit our educational module entitled, "Pain Management in Sickle Cell Disease" by clicking on the link provided on the Children's Hospital of Pittsburgh's Sickle Cell Program web-page at http://www.chp.edu/centers/03_hema_sickle.php. If you are a pediatric resident at Children's hospital of Pittsburgh you will be able to access this case from the PedsEd site under the heading, Pediatric Hematology.

Thank you. We look forward to your participation and feedback.

Sincerely,
Lakshmanan Krishnamurti, MD
Director, Comprehensive Hemoglobinopathy Program
Division of Pediatric Hematology/Oncology
Associate Professor of Pediatrics, University of Pittsburgh School of Medicine

APPENDIX D

CHP ALGORITHM FOR PAIN MANAGEMENT OF SCD PATIENT WITH PAIN IN THE ED

Sickle Cell Disease Vasocclusive Crisis ED Pain Management

Goal: Significant pain reduction in the first 1-2 hours

- **Begin Pain Management Immediately**
- ***Patients should be treated according to their individualized management plan (Refer to ED binder).***
- ***Maintain home medications throughout ED visit.***
- ***May hold short acting pain medications while pain is being controlled with IV pain medications.***
- ***If no current individualized management plan is available treat according to dosing guidelines below***

Supportive Care

- ✓Begin IV fluid 1-1.25 times maintenance
- ✓Begin antipruritics: (Per individualized management plan or CHP formulary)
- ✓Diphenhydramine (Benadryl) 0.1-.5 mg/kg or Hydroxyzine (Atarax) or 2-4 mg/kg/day in 6-8 doses.
- ✓Begin Antiemetic: Dose according to CHP formulary
  Ondansetron (Zofran)
  Proemethazine (Phenegran)
  Granisetron (Kytril)

Patient Assessment

- By self report determine characteristics, location and intensity of pain q 15-30 minutes.
- Assess pain with developmentally appropriate pain scale that the patient is familiar with and understands.
- Assess causes for pain, and change from baseline in spleen, O2 Sats, and mental status.

Pain related to SCD → **No** → Conduct complete workup to Determine etiology of pain

Yes

Treat based on characteristics of episode.

Assess for Sickle Cell Disease related complications.

Yes ←
- Acute chest syndrome
- Splenic sequestration
- Stroke
- Pneumococcal sepsis
- Dactylitis

→ **Admit to Hospital**

- Morphine 0.15 mg/kg IV bolus or
- Dilaudid 0.02 mg/kg bolus
- Ketorolac (Toradol) 0.5 mg/kg IV x 1 then: 0.5 mg/kg/dose IV q 6 hours (5day max) ***NOTE: limited by FDA, if used within past 30 days use ibuprofen.***

Morphine 0.05 mg/kg q 15-30 minutes until the pain is under control or as tolerated.

When pain is under control for 45-60 minutes, give an adequate dose of an effective oral opioid analgesic (refer to individualized pain plan or equianalgesic dosing table).

If pain not in control after 4 -6 hours after admission to ED, the patient is unable to take adequate fluids orally, if patient is in severe distress or if other complications suprevene

If significant pain persists or can not take adequate fluids.

Pain Relief maintained on oral medication for 1-2 hours

Discharge home with appropriate prescriptions and follow up appt. in Hematology Clinic in 1-2 weeks

CE # 210.02 Origination: 9/04 Revised:

This clinical guideline is a collaborative care plan and is not intended to construed or to serve as a standard of medical care. Rather, it is intended as a guideline to promote coordination and communication with respect to patient care and may be modified to meet individual care needs. For additional information contact the Department of Care Coordination at 412/692-7743

APPENDIX E

EVALUATION TOOL

Demographic Question 1
Job title

1. Resident
2. Fellow
3. Hematology Attending
4. Emergency Medicine Attending
5. Nurse Practitioner/Physician Assistant
6. Registered Nurse
7. Genetic Counselor/Social Worker
8. Psychiatrist/Psychologist

Demographic Question 2
Number of years since receiving highest degree

1. 0-3
2. 4-6
3. 7-9
4. 10-14
5. 15+

Evaluation Question 1
To what extent were the objectives of the educational activity achieved?

1. Very Low
2. Low
3. Moderate
4. High
5. Very High

Evaluation Question 2
To what extent were you satisfied with the overall quality of the educational activity?

1. Very Low

2. Low
3. Moderate
4. High
5. Very High

Evaluation Question 3
To what extent was the content of the educational activity relevant to your practice?

1. Very Low
2. Low
3. Moderate
4. High
5. Very High

Evaluation Question 4
To what extent do you feel that this type of web-based educational tool is useful?

1. Very Low
2. Low
3. Moderate
4. High
5. Very High

Evaluation Question 5
To what extent did the activity enhance your knowledge of treating pain in patients with Sickle cell disease?

1. Very Low
2. Low
3. Moderate
4. High
5. Very High (e5)

Evaluation Question 6
To what extent did the activity change the way you think about treating pain in patients with Sickle cell disease?

1. Very Low
2. Low
3. Moderate
4. High
5. Very High

Evaluation Question 7

To what extent will you make a change in your practice of treating pain in patients with Sickle cell disease as a result of your participation in this educational activity?

1. Very Low
2. Low
3. Moderate
4. High
5. Very High

Evaluation Question 8

To what extent did the activity help you to learn and understand the algorithm provided (Vasoocclusive Pain in Sickle Cell Disease)?

1. Very Low
2. Low
3. Moderate
4. High
5. Very High

Evaluation Question 9

Which of the following best describes the impact of this activity on how you will treat pain in patients with Sickle cell disease?

1. This activity will not change my behavior because I am currently treating pain in Sickle cell patients in a manner consistent with the information that was provided.
2. This activity will not change the way I treat patients with Sickle cell disease because I do not agree with the information provided.
3. I need more information before I change the way I treat pain in patients with Sickle cell disease.
4. I will immediately implement the information provided in the activity.

APPENDIX F

KNOWLEDGE QUESTIONS

Knowledge Question 1
A patient with Sickle cell disease presenting with fever and chest or abdominal pain should be evaluated for which of the following?

1. Sickle cell related complications such as acute chest syndrome or splenic sequestration.
2. Other medical or surgical problems such as appendicitis or peptic ulcer.
3. This patient is likely attempting to obtain more pain medications by complaining of chest pain.
4. 1 and 2 above.

Knowledge Question 2
What is the best way to determine initial dosing of pain medications for patients with Sickle cell disease?

1. All patients should receive IV fluids, oxygen and Ketorolac. If there is no response 2 hours after these medications have been administered, opiates may be considered.
2. Ask the patient what they took at home and continue to give them the same.
3. Follow the patient's established Individualized Management Plan or adhere to institutional guidelines for treating pain in patients with Sickle cell disease.
4. Dose at the low end of the range for age and body weight.

Knowledge Question 3
Which of the statements below correctly describes the goal of pain management and role of opioid use in patients with Sickle cell disease who present to the ED with pain from vasoocclusion?

1. Many patients with Sickle cell disease exhibit drug seeking behavior. It is important to minimize exposure to opioids to prevent drug addiction
2. Aggressive management of pain in a supportive environment can lead to relief of pain and obviate the need for hospitalization in a substantial proportion of patients

3. Aggressive use of opioids carries the risk of respiratory suppression and sudden death in patients with Sickle cell disease and should be discouraged
4. Patients with Sickle cell disease have a life long need for opioids and dosing should be restricted to the lower end of the range to prevent tachyphylaxis and drug dependence

Knowledge Question 4

At what intervals can pain medications be administered to a patient with Sickle cell disease presenting to the ED in a pain episode?

1. Since the duration of effect of morphine is 3-5 hours, doses can be repeated at that interval
2. Since the half life of morphine is 1-2 hours in children and 2-4 hours in adults, doses can be repeated at that interval
3. Any patient with Sickle cell disease coming to the ED should be given three fixed doses of Demerol at 2 hour intervals
4. Peak effect of morphine occurs in 20 minutes. Based on patient response, doses can be repeated at that interval

Knowledge Question 5

What is the best way to assess patient response to current management of pain?

1. The patient's self-report of efficacy of treatment
2. Patients with Sickle cell disease frequently exhibit drug seeking behavior. Treatment should be by a strict pain management contract regardless of clinical observation or patient report
3. Observation of the patient's activity and sleep pattern as reported by the nursing staff
4. Combination of clinical assessment, vital signs and laboratory signs of acute inflammation

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