Anesthesia Care for the Jehovah’s Witness Patient With Sickle Cell Disease
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Stem Case and Key Questions Content
A 12-year-old child is scheduled for tonsillectomy and adenoidectomy secondary to obstructive sleep apnea under general anesthesia. In the preoperative area, you meet the patient and her mother. Upon questioning of the mother and review of the consent form, you learn the parents are Jehovah’s Witnesses and demand their child be treated the same.

Are there any concerns with her medical history or parents’ religious beliefs?

What does it mean to be a Jehovah’s Witness?

Also, they explain she has sickle cell disease but “not the SS kind”, but SC hemoglobin. You further find the child had been recently hospitalized for acute chest syndrome. Currently, she feels fine. Her medications include: penicillin, folic acid, ranitidine, montelukast, fexofendadine, fluticasone, amitriptyline, and albuterol. She has no drug allergies.

How does one differentiate the hemoglobinopathies?

Is a blood transfusion a real possibility?

How will you approach the patient about the use of blood and blood products?

Would it be different if the child was an emancipated minor? or competent adult?

The physical examination reveals enlarged tonsils and clear rhinorhea. The lung sounds are equal but with a faint end expiratory wheeze. Her cardiac examination is unremarkable, except for tachycardia.

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Her laboratory examination includes a WBC -13.5, Hgb-11, Hct-30.3 and Plt ct- 471. Also, included in her preoperative laboratory work is an echo report from 6 months ago. The echo at the time shows trivial tricuspid regurgitation with a peak velocity of 2.4 meters per second, trivial mitral regurgitation, trivial pulmonary regurgitation and normal left ventricular end-diastolic dimension with qualitatively good left ventricular systolic function.

Are there any other studies needed before proceeding with the surgery? If so, which studies would you like to order and why.

Mom has had some time to think again about any heart problems and she remembers that her daughter had an evaluation by the cardiologist for sinus arrhythmia sometime last year and that it was normal.

Any concerns?

Any special considerations for preoperative or intraoperative management?

It was decided there would be no preoperative blood transfusion.

Are there any blood conservation therapies approved by Jehovah’s Witnesses?

In the operating room, the surgeon notes persistent oozing from the tonsillar bed. Suddenly, he notes brisk bleeding.

What happened? Your next moves? Blood transfusion? Are there any legal precedent to support a transfusion?

The bleeding is controlled and the patient stable. The child is transferred to the PACU and doing well.

You prepare to inform the parents of your decision to transfuse the child at this time.

Who should be involved in this discussion?

Thirty minutes later she complains of slight chest discomfort. She looks distressed and her oxygen saturation has fallen from 100% to 93% on nasal oxygen.

What are your concerns?
Jehovah’s Witnesses are a religious organization founded in Allegheny, Pennsylvania. It was founded by Charles Taze Russell in 1869. Initially known as Zion’s Watch Tower Tract Society, the name Jehovah's Witnesses was changed in 1931 based on a passage in Isaiah 43:10-11. The Blood Ban became into being around 1945. Jehovah’s Witnesses believe that the introduction of blood is not allowed based on several key passages of the bible. Genesis 9:3,4 3 Every moving animal that is alive may serve as food for YOU. As in the case of green vegetation, I do give it all to YOU. 4 Only flesh with its soul—its blood—YOU must not eat. Leviticus 17:14 14 For the soul of every sort of flesh is its blood by the soul in it. Consequently, I said to the sons of Israel: “YOU must not eat the blood of any sort of flesh, because the soul of every sort of flesh is its blood. Anyone eating it will be cut off.” Acts 15:28, 29 For the holy spirit and we ourselves have favored adding no further burden to YOU, except these necessary things, 29 to keep abstaining from things sacrificed to idols and from blood and from things strangled and from fornication. If YOU carefully keep yourselves from these things, YOU will prosper. Good health to YOU!”

In addition, the rejection of blood transfusions has been defended as being better medicine. Jehovah’s Witnesses support this by referencing large retrospective studies demonstrating Jehovah’s Witnesses at no greater risk of morbidity and mortality than other religious groups being treated for major trauma or from surgical procedures requiring a blood transfusion. In addition, they point out blood transfusions have been associated with a higher mortality, ICU length of stay and organ dysfunction. Also, studies have focused on defining the minimal hemoglobin concentration required to meet physiologic requirements in the human body. This value has often been quoted as >7 g/dL. However, some postulate the acute hemoglobin threshold for cardiovascular collapse may be as low as 3 g/dL to 5 g/dL.

The blood ban included initially whole blood and the components of blood. Consumption or intake of blood historically resulted in excommunication or organized communal shunning under Scriptural doctrine. However, a new directive was issued in 2000, replacing the Scriptural doctrine, which stated that by virtue of the transgressor accepting blood, the person had by defacto revoked their own membership.

Sickle cell disease is a genetic disorder with an incidence of 1 in 625 African-Americans. It is an autosomal recessive disorder. The predisposition of red blood cells to polymerize and sickle, results from the substitution of valine for glutamic acid in position 6 of the beta-globin chain of hemoglobin. Patients are chronically anemic as a result of the shortened half-life and hemolysis of erythrocytes. Oxygen delivery is partially compensated by the rightward shift of the P50 value for the oxyhemoglobin dissociation curve for sickle hemoglobin. The clinical manifestations are related to a chronic state of sickling and hemolysis with acute exacerbation affecting various organs by microvascular occlusion. The clinical hallmark is intermittent, recurrent, episodes of severe pain.
The diagnosis of sickle cell disease may be quickly performed by a sickle preparation or solubility test. However, these tests only detect the presence of sickle hemoglobin and are unable to identify a particular sickle hemoglobinopathy. The diagnosis is established by hemoglobin electrophoresis or more accurately with high performance liquid chromatography (HPLC). The results may show HbS constituting 80-95% of the circulating hemoglobin. The remaining hemoglobin is represented by HbF and HbA2. For newborn screening, many states have relied on the sensitivity of HPLC and isoelectric focusing to detect subtle hemoglobin variants. The implementation of a preoperative screening program has been proven to be ineffective. One variant form of sickle cell disease (HbSS) identified by hemoglobin electrophoresis is hemoglobin SC disease (HbSC). The proportion of HbS or HbC can vary between 45-50% with the remaining proportion consisting of HbF. The clinical severity is highly variable, but generally runs a milder course than individuals with HbSS. In a study conducted by Neumayr et al involving 92 patients with HbSC disease undergoing elective surgery, they concluded blood transfusions were beneficial for abdominal surgeries, but unnecessary for minor procedures such as myringotomies.

There are many types of sickle cell crises: vaso-occlusive, sequestration, aplastic and hemolytic. The most common form of crisis dealt with by anesthesiologists is vaso-occlusive. A vasoocclusive crisis results from the sickling of red blood cells at the microvascular level causing ischemia and infarction to the involved organ system, which is clinically manifested as pain. The most severe manifestations of vaso-occlusive crisis are acute chest syndrome, cerebrovascular accidents and priapism. Sequestration, aplastic and hemolytic crises reflect the acute pooling, lack of production and destruction of red blood cells, respectively. Many modalities of therapy have been targeted towards reducing the conditions conducive for the sickling process. The therapy is targeted to avoiding or preventing inciting factors, which interfere with oxygen delivery or increase oxygen consumption, such as severe anemia, hypovolemia, infection, and pain. Prophylactic antibiotics are administered to prevent serious infections as a result of a compromised immune system. Many children identified through newborn screening programs are placed on daily penicillin beginning at the age of 2 months until they are at least 5 years old. In addition, pneumococcus and meningococcus vaccinations are given along with routine childhood vaccinations. Painful crises are treated with a full spectrum of analgesics and techniques.

The most direct therapy for preventing or treating a sickle cell crisis has been to administer a blood transfusion to help optimize the flow or rheology of red blood cells and to increase oxygen delivery. Preoperative transfusion to reduce the blood’s viscosity and improve oxygen delivery has been a long-standing practice. However, oxygen delivery improves with hemoglobin only until the effects of viscosity again reduce flow and transport. There exist two methods to transfuse sickle cell patients needing blood: exchange transfusion and simple transfusion. In each instance, the patient receives normal erythrocytes, which can survive in vivo for 120 days compared to 5-15 days for sickle cells.
Traditionally, the goal has been to reduce the proportion of sickle cells to <30% and raise the hemoglobin to 10 gm/dl.

The results of a 1995 cooperative study comparing an “aggressive” versus “conservative” transfusion regimen showed no difference in perioperative complications. The aggressive group had blood transfusions to reduce the HbS to less than 30% and hemoglobin of 10 gm/dl and the conservative group had a simple blood transfusion to hemoglobin of 10 gm/dl. In addition, the conservative regimen had only half as many transfusion-associated complications.

In a follow up study in 1997 involving the same transfusion regimen and a non-transfused group, the same conclusions were drawn regarding the two transfusion groups; however, the highest incidence of sickle-related complications occurred in the non-transfused group. Similar findings have been reported by other investigators. In addition, they noted that the level of complexity of the operation should guide preoperative blood transfusions.

Preoperative sedation to minimize anxiety and/or help with placement of an intravenous catheter may prove very beneficial. Patient assessment for appropriateness of sedation should include the age, maturity, level of anxiety, and special needs. However, the use of anxiolytics must also be balanced with the avoidance of hypoventilation and hypoxemia.

Also, optimization for surgery includes intravenous fluid therapy to ensure adequate hydration prior to the induction of anesthesia. In a non-sickle cell patient, being denied liquids according to accepted anesthesia guidelines might not have a significant impact on the physiology of the patient. However, a patient with sickle cell disease becomes more at risk for a sickle crisis as he/she becomes more dehydrated and hemoconcentrated.

However, the most important aspect of taking care of a sickle cell patient, or any patient, is a thorough history and physical examination. Patients must be assessed for any acute illnesses, such as a respiratory infection. Sickle cell patients are more prone to infection because early in childhood most have lost the function of their spleen through multiple splenic infarcts and surgical splenectomy. Sickle cell patients are prophylactically treated with penicillin and vaccines to guard against encapsulated bacteria, such as pneumococcus.

Laboratory examinations must be ordered and reviewed. Many patients are chronically anemic and have a hemoglobin around 8-9 mg/dl. Many patients do not routinely receive a blood transfusion for simple operative procedures, such as bilateral myringotomies with tube insertion. In addition, cardiopulmonary, hepatic and renal function may need to be assessed, based on the patient’s history and physical examination.
A situation may arise where a child at risk may present for surgery and the parents do not know the status of the child or of themselves. The importance of determining whether a child has sickle cell disease is based on the increased risk related to anesthesia and surgery. If the parents sickle status can be obtained, one can determine the odds of the child’s sickle cell status based on Mendelian inheritance. This information can be combined with the hemoglobin and hematocrit results to help determine the next step. One might elect to have a sickle preparation ordered to avoid canceling the surgery. However, if the sickle preparation is positive, then the question of continuing with surgery becomes more critical. Based on previous studies of sickle cell patients, those having minor surgery may not be required to have a blood transfusion. Therefore, a delay in surgery for the results of a hemoglobin electrophoresis may not be necessary.

The intraoperative management of the patient should take into consideration the child’s underlying medical condition and the operative procedure. Sickle cell disease may affect multiple organ systems. Recurrent bouts of sickling leave the patient chronically anemic. The chronic anemia may result in cardiomegaly and high output cardiac failure. Respiratory insufficiency or failure may result from the chronic intermittent vaso-occlusion of pulmonary vessels. The child’s renal function may be impaired as reflected by the inability to concentrate urine. In addition, the child may have experienced a cerebrovascular accident. Also, hemochromatosis may add to the risk of anesthesia by affecting the same organ systems.

Ear, nose and throat surgery account for one-fifth of all procedures performed on sickle cell patients. Adenotonsillar hypertrophy is a common finding in pediatric sickle cell patients. In a study conducted by Waldron et al., they found obstructive sleep apnea was the most common indication for adenotonsillar surgery. The peak incidence of childhood obstructive sleep apnea occurs at the same time as painful crises and strokes in children. Patients having myringotomies were characterized as having a low risk for sickle cell-related events and receiving minor benefit from a blood transfusion. Also, they noted that patients with a history of pulmonary disease were at an increased risk for postoperative sickle cell-related events.

Additional considerations in caring for a sickle cell patient are more practical in nature. Intraoperatively, there may be a potential for rapid heat loss necessitating the maintenance of a warm environment with active warming of intravenous fluids and the patient. Positioning is very important. Positions, which may create venous stasis or a tourniquet effect, must be avoided. Also, the patient may become more prone to nerve injury with inadequate oxygen delivery. The use of cardiopulmonary bypass machines requires careful priming of the pump and maintenance of temperature.

The use of alternative anesthetic techniques and flexibility in case management is important when caring for the sickle cell patient. Especially challenging are those patients who present for elective
surgery without a recent evaluation or treatment from their hematologist. The actual technique may not be as important as avoiding episodes of hypotension, hypoxemia, hypothermia, venous stasis and acidosis. As discussed before, a blood transfusion may be unnecessary in minor procedures. Another consideration is when the operative procedure requires a bloodless field requiring a tourniquet. There have been numerous case reports concerning the use of tourniquets. In each instance care was directed at optimizing the patient’s hydration status, acid-base balance, oxygenation, reduction of sickle load and surgical stress. In some cases, the patient already possessed an advantage by having a relatively high percentage of fetal hemoglobin.

There have been many case precedents where patients with advance directives have died from a lack of a blood transfusion. The right to refuse medical treatment dates back to 1914 involving Schloendorff v. New York Hospital. Many Jehovah’s Witnesses carry a Durable Power of Attorney. This serves as an Advance Directive regarding their medical management. In addition, the card will list minor blood fractions and blood conservation technologies acceptable to them.

The acceptance of certain components of blood and blood salvaging techniques must be approached on an individual basis. Reformation movements within the Jehovah’s Witnesses have yielded the approval of the use of blood salvaging devices as long as they are in continuity with the patient’s circulation. In addition, autologous blood transfusions are acceptable as long as they are in continuity with the patient’s circulation. The use of blood substitutes has also been made a personal option. Other techniques are readily accepted such as intraoperative hypotension and isovolemic hemodilution, preoperative erythropoietin (EPO), use of recombinant Factor VII (rFVIIa). The health community in response to the growing membership of Jehovah’s Witnesses and the medico legal ramifications involved in caring for these patients, the medical community has developed programs to improve blood conservation programs. In addition, the Jehovah’s Witnesses have developed special Hospital Liaison Committees to help health care providers reach a mutually acceptable approach to medical management. Also, many institutions have developed a protocol for transferring care of patients, if the initial consulting physician is uncomfortable with respecting the wishes of the patient.

Involvement of the legal system to settle controversial debate is based on four guiding principles: preservation of life, prevention of suicide, protection of innocent third parties and preservation of the the ethical integrity of the medical profession. However, with the passage of time and the development of a reform movement, Jehovah’s Witnesses have softened their stance in regard to certain blood products, such as albumin, immune globulins, and hemophiliac preparations. In addition, autologous blood keep continuous with the patient’s circulation has been found acceptable by most Jehovah’s Witnesses.

This creates a legal conundrum suitable possibly for the invoking of the Principle of Parens Patriae.
This allows the state to intervene against an abusive or negligent parent, legal guardian or caretaker and to act as the parent of any child or individual who is in need of protection. This allows the state to order a blood transfusion to save the life of the child.

Mercy Hospital, Inc. v. Jackson in 1984, the Maryland Court of Special Appeals affirmed the denial of Mercy Hospital’s petition for appointment of a guardian for a pregnant Jehovah’s Witness in order to gain consent for a blood transfusion the medical staff deemed necessary to perform a Caesarean section. The Court of Special Appeals agreed with the trial judge that “a competent, pregnant adult does have the paramount right to refuse a blood transfusion in accordance with her religious beliefs, where such decision is made knowingly and voluntarily and will not endanger the delivery, survival or support of the fetus.”

The most referenced case law concerning parens patriae and a Jehovah’s Witness is Georgetown College v. Jones in 1963. A 25-year-old mother of a 7-month old child needs a blood transfusion after having lost two-thirds of her blood from a ruptured ulcer. The court cited the following: “The state, as parens patriae, will not allow a parent to abandon a child, and so it should not allow this most ultimate of voluntary abandonments. The patient had a responsibility to the community to care for her infant. thus the people had an interest in preserving the life of this mother.”

The postoperative anesthetic concerns are based on the child’s sickle cell disease. Continue postoperative care in avoiding hypotension, hypoxemia, hypothermia and acidosis. Provide supplemental oxygen and adequate analgesia, while avoiding respiratory depression. Maintain appropriate fluid therapy and, if the patient has a urinary catheter, ensure an adequate urine output. A very serious cause for rapid respiratory failure in the perioperative period is acute chest syndrome. The mortality rate is stated to be from 2% to 12% and accounts for 25% of deaths in sickle cell patients. The etiology is multifactorial, but ultimately related to the sequestration of sickled cells in small pulmonary vessels leading to pulmonary vaso-occlusion. The symptoms associated with acute chest syndrome are tachypnea, pleuritic pain, and cough. Radiological evaluation may show a new lobar infiltrate. Treatment is focused on stopping the sickling process and addressing the respiratory status of the patient.

In conclusion, the key to the successful management of a Jehovah’s Witness patient with sickle cell disease is planning. The more time one has to plan, the more prepared one will be to handle the uncertainties. A new concept which is being applied broadly to hospital patients is Bloodless Medicine Management. This is a multidisciplinary, multimodal approach to improving the patient’s blood indices. In addition, each Jehovah’s Witness patient is unique and may have a differing opinion as to what is an acceptable technique for blood conservation or blood constituent
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