

## SICKLE CELL DISEASE IN AN AIRMAN

### *First Known Certification via Allogenic Bone Marrow Transplant*

By BRUCE B. CHIEN, MD

**T**HE AIRMAN IS a 43-year-old male physician whose sibship included three of eight siblings who were homozygous for sickle cell disease (SSD). The other two siblings had already died due to complications of the disease. In 11/2009, because of continuous life difficulties presented by management of his SSD with hydroxyurea, he participated in a National Institutes of Health trial of bone marrow transplantation for SSD, the cohort report of which was reported in the *New England Journal of Medicine* by Hsieh M.M. et al., 361:14 2309-17.

His hepatitis C was treated with interferon and ribavirin prior to transplant. He did well and in February 2011 received his third-class medical certificate after initially being deferred.

In March of 2012 he developed a factor VIII antibody, and therefore hemophilia A (10% Factor VIII levels), manifest by compartmental bleeding that required fasciotomy in the right forearm. He self-grounded and immunosuppression was initiated transiently with rituximab and then with sirolimus 2 mg/daily. Factor VIII levels rebounded to 172 IU/dl, and he experienced a six-month period requiring no Factor 8 support and had no soft tissue bleeding. PTT was 30.3.

Three years after transplantation he has detectable haptoglobin, normal LDH, bilirubin, and reticulocyte counts; 96% of his CD14/15 cells and 29% of his CD3 cells are from his donor. He has split chimerism and had no hint of GVHD (graft vs. host disease). Quantitative hepatitis CRNA in October 2012 was negative.

The only organ system with a side effect was renal, and the airman continues to have a normal creatinine despite 20 mg/dl of proteinuria, likely due either to SSD or to sirolimus. His lipid profile was unremarkable. By surface echo he has been afflicted with none of the consequences of chronic iron overload. His life transfusional iron overload was treated with periodic phlebotomy, and his serum ferritin decreased from >4500 to 1292. He has retained normal LV function, HbA1c and thyroid functions.

On physical exam in October 2012, medications were sulfamethoxazole and trimethoprim (Bactrim) q.o.d. and sirolimus 2 mg qD. He was normotensive with a nonpalpable spleen. Protein was not detected on urine dipstick. A right forearm fasciotomy scar, healed by secondary intention, was present. The remainder of the exam was unremarkable.

After consultation with the Aerospace Medical Certification Division, he was authorized and was issued a third-class certificate for one year, contingent on annual current status reports. This is to our knowledge the first airman with homozygous sickle cell disease to achieve certification based on allogenic bone marrow transplantation.



*Dr. Chien is an aviation medical examiner and practices in Peoria, Illinois. He submitted this case report as the original examining aviation medical examiner.*

## MEDXPRESS VIDEO WINS TELLY AWARD

**T**HE CIVIL AEROSPACE Medical Institute's production of "MedXPress: It's Easy!" was selected to receive a 2013 Telly Award. Telly Awards are given to honor film and video productions, online video content, and TV commercials and programs. The MedXPress video was created to broadly socialize the new online system with potential users.

The video features an 11-year old aspiring pilot who shows his mother how easy it is to fill out his medical information online so that he will be prepared to get his medical certificate when he turns 16.

"We won a Bronze Telly in the Film/Video Category for informative TV programming," said Manager of Aerospace Medical Education Division **Brian Pinkston** (AAM-400). "We had a strong team that put the video together, led by **Alan Atkins** and **Laura Shepherd-Madsen** of AMI-700 [contract production organization]. They created the script and shot the video. **Susan Buriak**, an instructional system designer from AAM-400, managed the project and the team of AAM subject matter experts to ensure accuracy."

According to Dr. Pinkston, a second installment of the MedXPress video is in production and will be available soon; a video about aeronautical decision-making is being planned for production.



—Information provided by AVS Flyer

## CAMI TAKES HYPOXIA TRAINING TO HART

By J.R. BROWN

**T**HE CIVIL AEROSPACE Medical Institute announces the "maiden voyage" of its Hypoxia Awareness & Recognition Trainer (HART). This device is a direct descendent of CAMI's Portable Reduced Oxygen Training Enclosure (PROTE), which was another CAMI innovation. Each device allows participants to experience the effects of hypoxia by reducing the available oxygen by "scrubbing" it out. Air separators remove oxygen and replace it with inert nitrogen. By reducing the oxygen level from 20.95% to 7%, it effectively simulates an altitude of approximately 25,000 feet.

The HART is a normobaric "hypoxia training room" that utilizes the existing walls of the room to help contain nitrogen-rich air. This is another in a long line of CAMI innovations for the purpose of enhancing and promoting aviation safety.

Upon entry, subjects immediately begin oxygen desaturation, quickly feeling the effects of hypoxia. Within 5 minutes, they will feel several symptoms of hypoxia and will rapidly approach the time of useful consciousness. At the end of the 5-minute time limit, subjects don their oxygen masks and will have full recovery within seconds.

The advantage the HART and PROTE have over altitude chambers is that pressure changes do not become an issue. Individuals with head colds, upper respiratory infections, and seasonal allergies would have problems in the chamber because of possible barotrauma to ears and sinuses, but this, obviously, would not be an issue in a normobaric environment.

For further information on either the HART or PROTE, please contact the Airman Education Programs Team at CAMI by calling (405) 954-4837.



*Mr. Brown is a training instructor in CAMI's Aerospace Medical Education Division.*

## LETTERS TO THE EDITOR

### SLEEP APNEA AND FLYING

*Dear Editor,*

I was interested to see the excellent overview on the problem of obstructive sleep apnea (OSA) presented in the Feb. 2013 issue by Lt. Col. Allen ["Obstructive Sleep Apnea in a Pilot," FASMB, vol. 51, no. 1, p 8]. There continues to be a national, downward trend in scheduled sleep as well as the quality of our sleep. What was the exception is now the norm, and sleep deprivation is a given when dealing with patients and airmen. I suspect our addiction to electronic gizmos is partly to blame.

#### **Sleep Apnea and Aviation Safety**

As human beings, we have a near-zero ability to accurately self-assess our degree of impairment from fatigue. There is little agreement on how best to objectively measure impairment, as the MWT [maintenance of wakefulness test], which measures time to intentional sleep onset, is a much more accurate assessment of pathologic sleepiness, and that is what we really want to know.

Airmen need to be educated that being awake is not the "default mode" of "being." NREM [non-rapid eye movement] sleep is. When comfortable or bored, it is *normal* to fall asleep in 15 to 20 minutes. The greater the sleep debt, for ANY reason, the shorter the time to "powering down."

#### **Treatment and Medical Certification**

Many states, like Michigan (my state), have adopted a more rigorous screening process for CDL [commercial driver's license] exams than we do for our FAA exams. I understand the insurance industry has joined with the drivers' unions and came up with this compromise: Position therapy and dental appliances are not acceptable for treating OSA for CDL licenses in Michigan.

The concern regarding dental appliances and position therapy is that there is no way to confirm compliance or know when a pilot is "cramming" for an MWT test just before visiting an AME. Hence, we cannot ensure public safety the rest of the year. Also, there is very little information on compliance with dental devices. What little I could find suggests that it is actually worse than CPAP [continuous positive airway pressure]; about 50% at 30 months. Compliance with CPAP over 5 years is about 75%. Dental appliances can be very uncomfortable and cause dental problems, headaches, and TMJ [temporomandibular joint] disorders. The application of positive pressure support (CPAP) has gotten much better.

Most patients can successfully adapt to CPAP devices, and, referring to the airman in Dr. Allen's report, with an AHI 21.3, CPAP was the optimal treatment in this case, both for the airman's health and for aeromedical purposes.

Sleep apnea, effectively treated with CPAP, is the preferred method to effectively control the risk of daytime drowsiness. Adaptation to CPAP may require short-term use of medications, such as temazepam (Restoril). We include a warning for long-term soporific use, and a 72-hour warning [self-grounding] for occasional temazepam use in the process of adaptation to CPAP.

*Mark Ivey, MD*

[Spring Lake, Mich.]

*Col Mark Ivey, MD, is the California Air National Guard Chief of Flight Medicine, 129th Rescue Wing, Moffett Federal Air Field, Calif., and a senior AME. He is also in a solo private practice and board certified in Internal Medicine, Pulmonary, Critical Care, and Sleep Medicine. He has 2,400 hrs. Command pilot time in helicopters and is a CFI rotorcraft-helicopters with a commercial instrument rating.*

### PRINTING AFTER FORM SUBMISSION

*Dear Editor,*

In today's AME Bulletin ["MedXPress Lessons Learned" by Fred Tilton, MD, vol. 51, no. 1, p. 2], you say, "If you do not print an airman's certificate before you submit the exam, the system will not allow you to print a certificate. So, remember, print before you submit ...."

I ALWAYS submit before printing; I have never used Quick-Print. My method has worked reliably from the time MedXPress was first begun; in fact, I didn't even know of the Quick-Print option until I attended an AME seminar last year, as I always complete the exam myself with the airman next to me and have never needed the Quick-Print option.

I am confident that you mean that "Quickprint does not function after the exam has been submitted." In fact, once the certificate has been selected for display, it must be saved or printed right then, as we don't get a second chance. Because

of this, I have always saved a local copy of the certificate.pdf while it's still displayed, in the form, in a file under the directory D:\FAA\Certs+Forms\Nnnnnnnn.yr.mo.do.cert.pdf

Where Nnnnnnnn is the airman's medical chart number. This has saved my bacon many times with printer malfunction.

*Daniel L. Johnson, MD*

Menomonie, Wis.

*Dear Dr. Johnson,*

*Thank you for correctly pointing out that you can print certificates after submitting an exam. We clarified the statement you quoted on February 12—several days after it was first published—with these words: "So, remember to print before leaving that screen—LESSON 5."*

*Jana Weems*

Congressional Liaison

Aerospace Medical Certification Division