University of Nevada School of Medicine

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From Our Readers

Doctors Mark Hueftle, Colleen Morris, and Arrah Curry acknowledged appreciation of our efforts to record Nevada history of medicine.

Patricia D. Cafferata, Esq. writes, "Thanks for dedicating your article about Vietnam to Treat and the others who served there." It was brought to the editors' attention by Dr. Tom Brady that Dr. Jack Pershing (Carson City) was a Vietnam veteran. We regret that we were unable to find all of the names of Vietnam doctors from southern and rural Nevada. If you have such information please let us know so that we can acknowledge them in a future edition of *Greasewood Tablettes*.

THE SEED OF JOSEPH: A STORY OF GENETICS, FAITH, AND ENDOGAMY IN THE MOUNTAIN WEST (AN ETHICAL DILEMMA)

By Reuel Jake Measom (Winner of the 2014 Student History of A Disease Essay)

And cursed shall be the seed of him that mixeth with their seed; for they shall be cursed even with the same cursing. And the Lord spake it, and it was done. *The Book of Mormon*

In 1990 pediatric neurologist Dr. Theodore Tarby at Arizona State's Children's Rehabilitative Services, saw patients from the polygamist settlement of Colorado City, Arizona. Despite being well seasoned in practice, Dr. Tarby was reminded of the vastness of disease when a woman from Colorado City carried in her young son. Dr. Tarby quickly saw something was amiss and mental retardation was obvious. Even the untrained eye could see the physical abnormalities, particularly the boy's misshapen skull. Dr. Tarby was positive that this was not a case of cerebral palsy or Down syndrome and sent a urine specimen to Dr. Steve Goodman at the University of Colorado. Dr. Goodman discovered a highly unexpected condition so rare that

only 13 cases had been documented—a deficiency of an enzyme in the metabolic process of the Krebs Cycle.

The boy lacked fumarase, an essential enzyme for production of energy. The cause was unknown Dr. Tarby took a family history and discovered a daughter diagnosed with "cerebral palsy." He saw that she had the same condition as her brother, disproving that fumarase deficiency was a random chance mutation. A reality was soon recognized—the genes were inherited through selective breeding. These genes were located in the fundamentalist Mormon settlement of Colorado City, a place where blood lines were braided by arranged marriage and inbreeding.

John Yeates Barlow founded Colorado City, originally called Short Creek, in the 1920s. The settlement was the result of a schism within the Mormon Church following the 1890 Manifesto by Church President Wilford Woodruff that disbanded polygamy as a felony. The practice, originally taught by Mormon founder Joseph Smith, was considered an essential step in attaining the highest degree of heavenly glory, a place in the Celestial Kingdom. Government pressure and exclusion from statehood moved Mormon leaders to abolish the practice, but many considered polygamy to be mandated by God and felt that President Woodruff had fallen into sin. Thus, accordingly they sought solitude in the wilderness.

Joseph Smith Jessop followed John Y. Barlow as the community leader. Jessop, a deeply religious man and a patriarch within the Mormon priesthood, married his first wife, Martha Moore Yeates in 1889. They had 14 children and did not know that mixing their two bloodlines would cause fumarase deficiency. This became obvious when their 12th daughter married her second cousin, John Y. Barlow.

In the Fundamentalist Latter Day Saints (FLDS) a prophet governs the community, and important decisions go through him, including marriages. To maintain "pure bloodlines" approximately 80% of the community is related to Joseph S. Jessop or John Y. Barlow. Benjamin Bistline, a historian and former member of the Short Creek polygamist community, labels the issue "religious totalitarianism." Despite the discovery of fumarase deficiency and repeated warnings that the practice of endogamy will

propagate the problem, the community did not take heed.

In 1937 Hans Adolf Krebs established and formalized the concept of the citric acid cyclealso known as the tricarboxylic acid (TCA) cycle or the Krebs Cycle—as a cornerstone in human's metabolic processes. Its role in creating useful coenzymes from carbon-based molecules is crucial for life. A paper in the The Mitochondrion states, "This pathway is so crucial to the metabolism of living cells that any significant defect is incompatible with life." A few years before Dr. Tarby stumbled on fumarase deficiency, a defect existing in the TCA cycle was believed to be an impossibility and incompatible with life.

Donald Whelan described the condition in 1983 when he and his colleagues reported on 2 adult siblings with fumaric aciduria and mental retardation. They discounted an inborn error of the fumarase enzyme, citing that the only place fumarase has a function is in the TCA cycle, and any error would be disastrous. The enzyme defect was not recognized until 1986 when Arthur Zinn reported an infant male with severe early onset encephalopathy. Zinn became convinced that the boy's condition was fumarase deficiency.

Doctors Tarby, Kirk Aleck, and a team of physicians from St. Joseph's Children Hospital in Phoenix went to Colorado City. The result was a landmark study on fumarase deficiency and the largest documented cluster in the world. They reported 8 cases and thought there might be more. The disease is inherited in an autosomal recessive fashion;

hence both parents possessing the gene increased the likelihood of its occurrence. Aleck and the others describe the effects of the disease in detail. The disease causes seizures, hypotonia, and early onset encephalopathy, but the key to diagnosis is excessive urinary fumaric acid.

The researchers traced the defective gene to Joseph S. Jessop and wife Martha. They discovered the extent of the gene's distribution and demonstrated that most of the residents are related to the patriarch. They discussed the situation with church leaders and explained that intermarriage produced children with disabling abnormalities. Their warnings fell on deaf ears and town members were convinced that it was in the water or a result of older women having babies. Historian Bistline stated, "It is something that is simply not talked about." It is a taboo extending from the belief that physical maladies are a manifestation of God's indignation towards sin. Despite the polygamists' indignation, Doctors Tarby and Aleck convened a meeting and laid out what it would take to eradicate the condition. These measures would require the genetic screening of members and the prohibition of marriage between carriers of the gene. It also meant that families showing the condition should cease having more children. The FLDS did not accept these conditions.

The community was not willing to make changes because the condition is uncommon. Bistline, who married into a mix of the Barlow and Jessop line, described two of his nieces with the disorder. He stated that he

was lucky that none of his children possessed the malady. He felt his progeny were able to "over power" the Jessop line. He alluded to the notion that there is a negative connotation with the Jessop line in conjunction with the illness. Whether the polygamist community believes the condition to be due to past transgression is unknown, but some believe that the seed of Joseph S. Jessop is cursed. What is known in the scientific community is that as long as intermarrying continues the condition will grow.

When Dr. Tarby conducted the study, FLDS prophet Warren Jeffs, who had not been seen since 2003 and is now incarcerated, was part of the community. To prevent the children from being photographed the doctors conducted the clinic in Arizona, but they feared the distance also would inhibit genetic counseling. Colorado City's religious influence effects the medical community and government officials in protecting future progeny. People have religious freedom rights that can cause propagation of a terrible illness. Sadly, obedience to their beliefs, such as living in isolation, not marrying outside of the faith, and avoiding worldly teachings have brought this genetic curse. The problem is not going away.

In December 2013 U.S. District Court Judge Clark Waddoups essentially decriminalized polygamy in Utah. This ruling changes the dynamic of the polygamy and the potential spread of fumarase deficiency. Where previously the government had the potential to intervene on behalf of the children from polygamist marriages, the new ruling gives legitimacy to the practice. Judge Bistline noted that since the discovery of the condition there have been schisms within the FLDS

community resulting in a higher likelihood of inbreeding. This could result in an increased opportunity for propagation of the defective gene.

As there is no cure for the condition, and the FLDS show

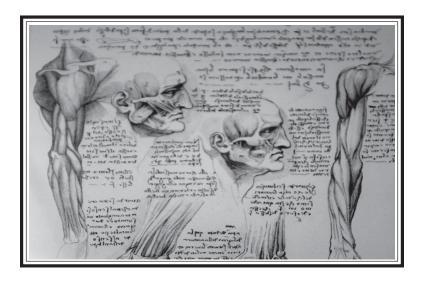
no signs of altering their religious practices, an ethical question is raised. What is the role of the scientific community and government to protect the unborn and prevent practices known to cause genetic diseases?

Dr. Hugh Collett Family Donates Leonardo da Vinci Anatomical Etchings To The School Of Medicine

The Collett family donated in June 2015 anatomical etchings by Leonardo da Vinci to the University of Nevada School of Medicine. They are now displayed in Savitt Library at the entrance to the History of Medicine Library. Elko's Dr. George A. Collett collected the etchings over 80 years ago. Leonardo da Vinci made the muscle-skeletal drawings around 1510, and in 1796, John Chamberlaine published his Imitations of Original Designs by Leonardo da Vinci, with etchings by Francesco Bartolozzi. The original etchings are on display at the Royal Library in Windsor Castle.

No man made such a tremendous impact upon the city of Elko within the short space of seven years, as did Dr. George A. Collett, who came to Elko 11 December 1946 after practicing in Crawfordsville, Indiana. He died suddenly at 63 years of age after practicing in Elko for 7 years. His dreams for a future medical center were promoted under his dynamic leadership. He was also involved in civic affairs and was named president of the Elko Chamber of Commerce.

Elko's medical heritage was primarily due to the standard of medical and surgical care of the earlier partnership of Dr. A. J. Hood, Dr. Charles Secor, and Dr. R. P. Roantree. They had succeeded not only in providing excellent medical and surgical care, but they had done it in such a professional manner that the medical atmosphere in Elko was one



of cooperation and helpfulness among all of the physicians. Elko's doctors organized the second medical clinic in Nevada four years after the Las Vegas Clinic. Doctors George A. Collett, R. P. Roantree, Leslie A. Moren and Dale Hadfield established the Elko Clinic in 1946. Retired Doctors A. J. Hood and Charles Secor were associates and had their offices in the clinic building.

In addition to guiding the Elko Clinic, George Collett served as a member of the Nevada Board of Medical Examiners. At the time of his death he was second vice president of the Nevada Medical State Association and on the Nevada State Board of Education. His son Hugh continued his legacy in civic and medical circles. Hugh recently received the Distinguished Nevadan Award from the Nevada Board of Regents.



Dr. Hugh Collett and Dr. Sohn Photo by Ann McMillin

Dr. Phil Usera Appointed Co-Editor

Greasewood Tablettes is adding a new coeditor. Dr. Phil Usera, Assistant
Professor in the Department of
Pathology at UNSOM. He will
contribute articles and assume
responsibility in future issues. He is also

investigating the naming of a Las Vegas editor. Although *Greasewood Tablettes* has printed articles on history of medicine in Las Vegas, we wish to have more input from a doctor living in southern Nevada.

A Sad Day, Good Luck and Farewell to Teresa Garrison

We are sad to report that Teresa Garrison our Associate Editor since 2003 will be moving to

Texas. She not only has typed all of the issues for the past twelve years, but she designed *Greasewood's* present format. Teresa's guidance contributed greatly to our success. Her loyalty to Nevada's History of Medicine continued after she retired in September 2013 from the School of Medicine's Pathology Department. Since then she has continued to format *Greasewood Tablettes*. We send our best wishes to her and Tom, who will be retiring as Sparks' Fire Chief.

GREASEWOOD TABLETTES © is a quarterly publication of the Department of Pathology, Great Basin History of Medicine Division, University of Nevada School of Medicine. Doctors Anton P. Sohn, Robert Daugherty and Phil Usera are co-editors, Lynda D. McLellan is our production manager, Dr. Marcus Erling is publisher, and Kristin Sohn Fermoile is copy editor. The newsletter is printed by the Department of Pathology. The cost of publication is paid for by a grant from the Pathology Department, School of Medicine. The editor solicits any items of interest for publication. Suggestions, corrections and comments are welcome. Please feel free to email us at antonps@gbis.com or write us at Department of Pathology/0350, University of Nevada School of Medicine, Reno, NV, 89557. The name GREASEWOOD TABLETTES © is derived from the greasewood plant or creosote bush, a plant that was used by Native Nevadans for medicinal purposes. It is still the subject of pharmacological research today.