



SPRING
2013

Happy Spring

With all the snow still left on the ground, I am annoyed that the Ground Hog lied to us! Hope that warm and sunny days will arrive soon.

The Education weekend is planned for May 24/25 and must congratulate Lisa and her committee for all the hard work they have been doing to make this a great weekend. The registration page will open on April 1, hope that many of Region Members can attend. Just click on the Education link on Region IX website to register, Region IX accepts, MasterCard, Visa and Paypal.

The new awards are ready for all members to apply, and the vendors have money to give away, so please do not be shy and apply. We also need to keep our Awards committee busy.

I did not receive any submissions for this newsletter, so I am submitting one that I completed for my 2013 CMLTA yearly learning plan, hope you find the article interesting.

Ann Lynde
Region IX Director

Spring is nature's way of saying, "Let's party!"
Robin Williams

NSH Summer and Annual Convention Symposium

Region IX members interested in attending the NSH Summer Symposium, it will take place at the Planet Hollywood in Las Vegas on June 17-18, information can be found at www.nsh.org

Chart your Course to the 2013 NSH Convention/Symposium in Providence, Rhode Island
September 20- 25

Watch for more information from NSH regarding registration!

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Future Dates
40th Annual Symposium/Convention
August 21-27, 2014
Austin, TX

41st Annual Symposium/Convention
August 28-September 2,
2015
Washington, DC

Human Hand Walkers

A few years ago I was intrigued when I watch a program on Nova about the Human Hand Walkers that I purchased the video from Nova/PBS. In my research I also found a family of three sons in Iraq that also never stood up. I will start with the family from Turkey.

At about one year old, human infants typically take a step that distinguishes them from – and raises them above – all other primates. Not so with five affected siblings belong to a family of nineteen children who live in a village in Southern Turkey. Their existence was well known in their neighbourhood, and they had several times been the subject of news reports in the local media. Of sixteen surviving children, ten are unaffected and walk in a normal biped way, five (four females, one male, aged now from 27 to 43) walk quadrupedally, and one (male, age 41) has cerebellar symptoms but walks upright. Another male child also walked quadrupedally but died aged 5.

(The pattern of inheritance strongly suggests that the abnormal brain development is the result of an autosomal recessive mutant gene, presumably present in both parents, and combined homozygously in a proportion of the children (expected 25%, observed 37%). The Dysequilibrium Syndrome, in the Hutterite population of North America, has been shown to be just such an autosomal recessive disorder.)

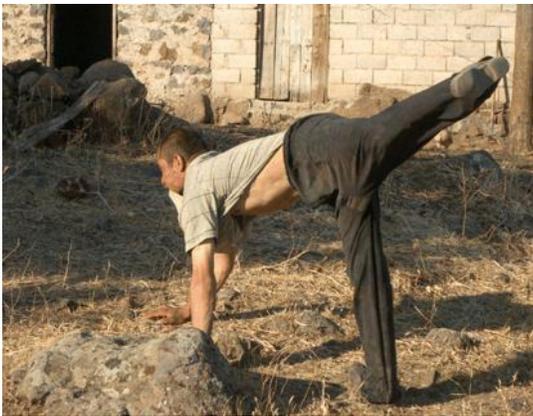
Behavioural observations were made at their home and farmyard in, and in a hospital.

When they are at rest the quadrupeds either sit upright or squat on their haunches. They are able to stand upright, without support, if they concentrate on doing so, and they lock their knees in a normal human way; but they become unsteady if they try to walk bipedally, and soon go down onto their hands. Their preferred form of locomotion, even when climbing or descending steps, is on all fours.



They move in this way fluently and effectively, and seemingly without discomfort. This contrasts markedly with normal adult humans who find such a gait – if and when they try it – tiring and uncomfortable even after practicing. They step with their hands and feet in both diagonal and lateral sequence gaits. The hands are placed palm down, with the weight taken on the wrists and lower ulnar area of the palm. The fingers are either arched so that the finger-tips make light contact with the ground or else are raised entirely clear (so that they can in fact hold objects while walking). The arms and also the back legs are nearly straight and stiff during the stance phase, with the bottom raised high, and the backbone remaining straight throughout the gait. Even as they step with their feet, the knees show relatively little flexion, so that the leg movements actually resemble those of normal human bipedal locomotion. The females splay their back legs apart, the male, however, who is the strongest and most active of the five, plants his feet closely together.

The local villagers laugh at and tease them. Because of this, the females tend to stay close to the house, but the male sometimes wanders for several kilometres. He helps raise money for his family by collecting cans and bottles, which he carries home in a pouch made from his shirt, held by his teeth. He is remarkably agile. We watched him moving easily across rough terrain in search of collectibles. While he searched ahead, his hands anticipated the contours of the rocks, so that he placed them deftly without looking down. He was able to run ahead of us, carrying his mouth bag – while at the same time, to show off, he kicked one of his legs in the air.

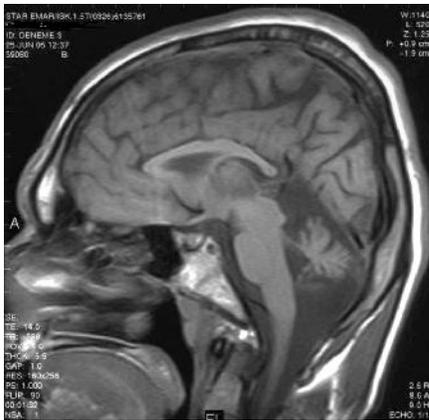


All their hands are heavily callused in the area of the ulnar palm and wrist where they make contact with the ground. But their fingers have been protected by being raised off the ground and show little extra wear. Even though their hands are used for locomotion, they retain considerable dexterity. They are slower than normal at manual tasks, and apparently must give the task their full attention. Nonetheless, the females are able to thread needles and do delicate crochet-work.



They have not attended school and have had limited exposure to the world beyond their home compound. They are mentally retarded, to differing degrees. They do not speak a human language and make only animal-like noises, they can all speak and understand Kurdish well enough to communicate within their own family and have reasonably good interpersonal skills.

All five show signs of cerebellar dysfunction including: intention tremor, inability to execute rapidly alternating movements particularly of the limbs, lack of coordination of movement typified by under- or overshooting the intended position and involuntary rhythmic eye movement, with the eyes moving quickly in one direction, and then slowly in the other. However, the cerebellar signs are relatively mild, and they are no more pronounced in the quadrupeds than in the one affected brother who walks bipedally. MRI scans of the brain confirm, as expected, that all the affected individuals have cerebellar hypoplasia, particularly affecting the vermis. The cerebral cortex also shows mild atrophy and the corpus callosum is reduced.



Iraq

The three brothers – Ali, 41, Abbas, 36, and Qusay, 34 - what they want most in life they all give the same poignant answer: to walk and get married. Ulaywi (their father) is a humble man, a former school cleaner who farms a little land in the village of al-Nakhella. He married his first cousin, Hassna, and they have had ten children – five boys and five girls – whom they raised in a tiny two-roomed house. Three sons and the eldest daughter, Ahlaam, 44, are hand-walkers. Ulaywi's youngest daughter has Down's syndrome. Ulaywi says that the four hand-walkers never crawled on their knees like normal babies. From infancy they used what is called the "bear crawl" with their legs held straight. He realized when Ahlaam was three that something was wrong and tried to teach his children to walk on two legs, using crutches and other supports, but without success. He went to many doctors and hospitals, including the best in Baghdad, but they sent him away saying the problem was congenital, or the result of inbreeding, or simply the will of Allah. Eventually he resigned himself to their condition.

The brothers never received an education, however. All three are a little slow mentally and slur their speech, and each was sent home after just a few days at the village school.

The brothers walk on all fours in identical fashion. They all say that walking upright hurts their lower backs, and that they lack balance.

Like the family in Turkey, they put their weight on the heavily calloused heels of their palms, sometimes topple over when sitting, find stairs hard, and have to go down backwards.



But in other ways the siblings are quite different. Qusay is also the best on two legs. He walks stiffly, with his legs splayed out, but can cover 50 yards or more. He practices regularly with a stick and still believes that one day he will be able to walk normally.



Qusay is also the heaviest of the trio,



and the least good on all fours. The three brothers recently embarked on a 60km pilgrimage from their village to the holy city of Karbala. Qusay managed 40km, but, incredibly, Ali and Abbas completed the entire journey in 30 hours.

It appears obvious that the problem is congenital, but there are two other families in the same village, neither with any familial connection, with offspring that hand-walk. The oldest son is a hand-walker and cannot talk, of the four other siblings, two are healthy and two have learning difficulties.

I could not find any articles on the second the family.

The discovery of all quadrupedal siblings therefore presents a considerable theoretical challenge. Whatever else may be going on, the explanation must presumably begin with the dysfunction of the cerebellum. Without the cerebellar problem, these individuals would surely have learned to walk bipedally, like their unaffected siblings. Even with the cerebellar problem, it might have been expected they would have achieved bipedal walking in some manner. The capacity for walking upright is highly resilient in human beings. In fact humans typically remain bipedal in the face of much greater obstacles to balance. Some suggestive evidence. (i) The mothers of the quadrupeds mentioned the normal ones as well as the quadrupeds -- were in fact bear-crawlers as infants. (ii) After the first such child, there were other quadrupedal children to serve as models. (iii) The fathers regarded their "crippled children" as a gift, sent by God or simply the will of Allah to provide them with the opportunity to prove their capacity to love and take care of them.

Go to this link to watch the trailer from PBS: www.pbs.org/wgbh/nova/preview/w_3317_220.html

References: PBS/Nova
London School of Economics and Political Science
The Times

NSH Region IX would like to extend thanks to Leica Microsystems for their continued sponsorship of
The Cutting Edge Newsletter



2013
NSH Teleconference/Webinar

March 27 : 1 pm ET:
Training for the Revised Hazard Communication Standard (HCS)

April 24 : 1 pm ET
MOHS: What's It All About

May 22: 1 pm ET
Emotional Intelligence

June 26: 1 pm
Identify Histology Look-Alike

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Living up to Life



Br
Breast Pathology

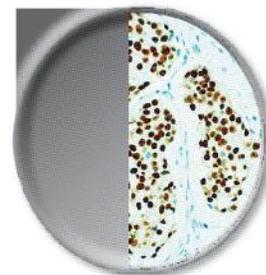
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