

COMMENT ON JACOB SHEEP AND TAY-SACHS DISEASE: A CONTRIBUTION TO CELEBRATE, NOT A PROBLEM TO FEAR.

The American Livestock Breeds Conservancy (ALBC) recently published an article summarizing the use of Jacob sheep for research on human Tay-Sachs disease (TSD) (Nov/Dec 2011). This is an amazing contribution to medical science – one that arose primarily from the efforts of Jacob breeders Fred and Joan Horak, who co-authored the article along with research scientists. The Horaks' decade long, open collaboration with the scientific community is one that we should all emulate, both as livestock breeders and as caring people.

Unfortunately, however, this article inaccurately portrays the Tay-Sachs mutation as a common one in Jacob sheep, suggesting that it is a defect that has been overlooked by Jacob breeders. This misrepresentation of the work done in identifying carriers could, ironically, discourage people from breeding Jacobs – even though it was conservation of the breed that led to the Tay-Sachs research.

Please consider the following:

- The initial primary purpose of testing Jacobs for TSD was to identify additional sheep that could be used for Tay-Sachs research. Testing was initially focused in flocks known to be affected or where symptoms of the disease had been seen – including the Horak flock where breeding to intentionally produce more carriers was done. While a secondary purpose was to identify carriers for breeding purposes, testing was voluntary, and was not systematically carried out in a large number of flocks.
- The experience of the Horaks is not typical – there is no evidence that TSD is a common problem in Jacob sheep. Reportedly, almost 600 Jacobs were tested, and 150 carriers found. However, it is important to note that this represents the percent of carriers only in the group tested – which included only a small number of flocks, and does not represent how common the gene is in the general population. Again, the testing was carried out with a primary goal of finding carriers for research.

There are easily 10,000 Jacobs in existence. The 150 carriers that were identified are only a small percentage of this total population (about 1.5%). While we don't know the % of carriers in the untested population, the sampling that was done is far from random, and cannot be applied to the population as a whole.

According to a January 2012 report from the National Tay-Sachs and Allied Diseases Association research report, testing was done in only 10 flocks. Carriers were found in 90% of the flocks tested – that is, in only 9 flocks. Again, it was done primarily where the disease was suspected due to the loss of lambs and occurrence of symptoms of TSD. The most recent JSBA Flock Book lists several hundred registered flock prefixes, and over 150 current members. JSC lists 25 member farms (some of whom are also JSBA members). Most breeders, like us, have never seen symptoms of TSD, and did not see the need to test.

- The ALBC article suggests a scientific concern that breeders have failed for 40 years to observe or report the death of lambs at 3 to 6 months from TSD. In fact, very few Jacobs were being bred 40 years ago. That aside, the lack of reporting of lamb death from TSD simply reflects that fact that most people have not seen symptoms of TSD. The discovery of this problem in the Horak flock was widely reported, and with free testing one would assume that those who were concerned about symptoms in their flocks followed through and identified carriers. For most of us, though, this was simply a non-issue.
- If the TSD mutation is uncommon, it is unlikely that many lambs will be affected (that is, both parents must be carriers), and affected lambs do not survive to reproduce. In the offspring of a carrier bred to a non-carrier, statistically only half of the next generation would be carriers. And in the second generation, only a quarter of the offspring would be predicted to be carriers, in the third generation an eighth, and so on. In this way, the incidence of the gene will tend to be reduced, even though

one of the early foundation rams (Turner 183K) apparently introduced the gene to a number of early offspring. In those unlucky few flocks that included multiple carriers, the symptoms will eventually be identified in affected lambs, as they were in the Horak flock.

The relative rarity of the TSD mutation means that it should not be a concern among North American Jacob breeders. It is not an “identifying trait” or a significant weakness in the breed. Neither current nor potential breeders should be concerned about purchase of breeding stock from flocks where testing had identified carriers, or from flocks that are without any symptoms of TSD.

As Jacob breeders, we have every reason to celebrate the contribution of the breed to human health, and no reason to portray Jacob sheep, or Jacob breeders, in a negative light as an outcome of this research. Jacobs remain a hardy, healthy, and fascinating breed – and now a few of them get to wear a “Hero” badge as well.

Contributed by—

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The Jacob Sheep Breeders Association registers 300-500 Jacobs per year. Presumably an equal number are not registered or registered only through JSC. If one assumes an average age of 10 years for existing sheep, there are 10,000 Jacobs in existence.

The ALBC article indicates that ultimately 16 breeders participated in the project.

Sweetgrass Jacobs has been breeding Jacob sheep since 1987, producing an estimated 500 – 750 Jacob lambs during that time. Sweetgrass has never had a lamb die with TS symptoms, although we have stock that go back to the Turner 183K ram. We did test one ram lamb for a buyer who requested the test; that lamb was negative.

