

BARC's Anna-Sophie von Ungarn -- "Sophie Tucker"
Expert at Cuddling in Bed and Survivor of
Puppy Mills, Dog Auctions, Liver shunt, Mast Cell Cancer, & Various Research Studies

Drs. Nancy Melone & Timothy McGuire, Sophie's Adoptive Family

"Sophie Tucker" was born in an Amish puppy mill not far from Walnut Creek, Ohio. She never made it to the Buckeye Dog Auction because at three months she weighed only 13 lbs. By contrast, her brother weighed 32 lbs. Clearly, something was wrong. That something was a genetic defect -- Sophie inherited the gene for intrahepatic portosystem (liver) shunts (IHPSS) from both of her (unaffected, but gene-carrying) carrier parents. While her brother was not affected (*i.e.*, he appeared to be normal), there is on average a 50% probability that he inherited one gene from either his mother or father. With one gene, he would be an unaffected carrier of shunts like his parents. Fortunately, when he was rescued, like Sophie, he was sterilized.

The mode of inheritance for IHPSS in Bernese Mountain Dogs is thought by University of Utrecht (Netherlands) geneticists to be through an autosomal recessive gene, similar to vWD in Bernese. If both parents are unaffected carriers of the shunt defect (like Sophie's mom and dad), they are likely to produce a litter with on average 25% shunt-affected puppies (like Sophie); on average 50% shunt-gene carrier puppies (like Sophie's mom and dad), and on average 25% genetically normal puppies. While we can generally identify an affected puppy (*i.e.*, one that has a shunt), ***we cannot differentiate between the on average 50% (unaffected) carriers of the shunt gene and the on average 25% genetically normal dogs*** produced from two (unaffected) shunt-gene-carrying parents. ***Currently (3/07) we have no genetic test to identify (unaffected) carriers of the shunt gene.*** We need to develop one before it is too late.

What do YOU know for sure when YOU hear about a litter containing a shunt puppy? You know that ***BOTH PARENTS*** of that litter are ***carriers of the shunt gene*** and ***every puppy*** from this pairing ***has on average a 75% probability of being either an affected shunt puppy or an (unaffected) shunt carrier.*** Experts advise that RESPONSIBLE BREEDERS would remove BOTH PARENTS from breeding programs. Even if one of the carrier parents is later bred to a normal (non-carrier) dog, every puppy produced from the genetically normal dog and the shunt-gene carrier dog will have on average a 50% probability of being an unaffected carrier of shunts. ***The take away? If you are looking for a breeding prospect from a litter with two carriers or even one carrier as parents, the message is clear -- keep looking and pray that there are a lot of honest breeders out there who keep good records of their progeny and necropsy all "failed puppies" to be sure they did not die of IHPSS.***

Pedigree research indicates that we have IHPSS carriers among the very best examples of our breed, those most likely to reproduce. Hence, we cannot turn our backs on this or we will find a breed rampant with carriers in only a few years. As a breed, our goal must be to eliminate the production of shunt puppies and unaffected carriers. This can be accomplished most quickly by developing a genetic test to screen for them. IHPSS gene isolation and genetic tests are developed with DNA/Blood drawn from: 1. dogs that have produced litters in which there was a shunt puppy and 2. the shunt and remaining puppies in that litter. Let's face it. Not every breeder has what it takes to do the right thing. It takes a person who values the welfare of the breed over the reputation of any single dog, regardless of how wonderful that dog is. For owners of "normal" and affected puppies from known shunt litters, you, too, can contribute DNA for the study. Everything is confidential, including your participation.

Please participate. **No single dog is worth sacrificing an entire breed. Please, do the right thing and help us convince others to do so, too.** Contact Nancy Melone at nmelone@nauticom.net.

Follow up -- Sophie underwent experimental minimally invasive surgery at University of Pennsylvania to close the shunt. Sophie is off all shunt medications and eats a normal diet, but she is not a normal puppy. Although she has a zest or life, she is a slow learner -- probably because of the neurotoxins that once bathed her brain.

A Canine Good Citizen certificate is her aspiration. Last year, Sophie Tucker was diagnosed with a grade II, mast cell tumor. It was removed surgically. In February, she celebrated her first year - cancer free. **Sophie Tucker plans to have a really good time at her 3-year birthday party on June 1st.**

“SOPHIE TUCKER” BARCs ANNA-SOPHIA von UNGARN

AKC WS09285001 and Berner Garde Dog ID 22587



Pictured at time of rescue at 3 months and 12 lbs suffering from normally fatal intrahepatic portosystemic (liver) shunt (IHPSS), hemorrhagic diarrhea, Coccidia (internal parasite) and Campylobacter (bacterial infection).