Björnstad syndrome is an extremely rare inherited disorder characterized by the presence of abnormally flattened, twisted hair shafts (pili torti) and, in most cases, deafness (sensorineural hearing loss). Hearing loss typically affects both ears (bilateral). Individuals with this disorder usually have dry, fragile, lusterless, and/or coarse scalp hair as well as areas of patchy hair loss (alopecia). Both autosomal dominant and recessive inheritance have been reported in the medical literature. Björnstad syndrome is an extremely rare disorder that was first described in 1965. In theory, it affects males and females in equal numbers. However, in observed cases, more females than males have been identified. More than 30 cases have been reported in the medical literature. The diagnosis of Björnstad Syndrome may be suspected by the finding of twisted hair (i.e., pili torti), which may be obvious at birth. The diagnosis is confirmed by examination of hair shafts from affected individuals under an electron microscope, demonstrating characteristic twisting of the hair shafts at regular intervals. Since the presence of this hair abnormality is suggestive of Björnstad Syndrome, all infants with this finding should be evaluated for possible nerve deafness. Sensorineural deafness may be confirmed through a variety of specialized hearing (auditory) tests.