BAP1 tumor predisposition syndrome is an inherited disorder that increases the risk of a variety of cancerous (malignant) and noncancerous (benign) tumors, most commonly certain types of tumors that occur in the skin, eyes, kidneys, and the tissue that lines the chest, abdomen, and the outer surface of the internal organs (the mesothelium). Affected individuals can develop one or more types of tumor, and affected members of the same family can have different types.

Some people with BAP1 tumor predisposition syndrome develop growths in the skin known as atypical Spitz tumors. People with this syndrome may have more than one of these tumors, and they can have dozens. Atypical Spitz tumors are generally considered benign, although it is unclear if they can become cancerous. Skin cancers are also associated with BAP1 tumor predisposition syndrome, including cutaneous melanoma and basal cell carcinoma.