

## Turner Syndrome Ovarian Removal Statement

Turner syndrome (TS) is a chromosomal abnormality that occurs in approximately 1 out of every 2,000 live births, affecting all ethnicities and nationalities. Gonadal dysgenesis is a finding known to be associated with TS.

Per the Clinical Guidelines for the Care of Girls and Women with Turner syndrome published in 2017, “The rate of gonadoblastoma among TS patients with Y chromosome sequences that were detected by PCR or FISH varied from 4 to 60% in 14 studies, and data on the long-term outcome of this cohort are incomplete. Putting all data together, approximately 10% may develop a gonadoblastoma, although there is considerable variation in risk estimates, possibly related to methodology, sample size and potential selection bias.”

In a 2018 study where nine patients, it was found that patients who presented with malignant ovarian germ cell tumors in the context of gonadal dysplasia had “higher rates of events and death” than those with normal gonads.” The authors recommended early bilateral gonadectomy for patients known to have gonadal dysplasia with Y chromosomal material. They also stated that, “In contrast to those with pure dysgerminoma, these patients may represent a high-risk group that requires a more aggressive chemotherapy regimen.”

**The current recommendation per these guidelines is gonadectomy in all female individuals with Y chromosome material identified on standard karyotyping. This should be a considered coverage by all insurances in light of the risks of not performing a gonadectomy in these TS girls.**

Gravholt CH, Andersen NH, Conway GS, Dekkers OM, Geffner ME, Klein KO, Lin AE, Mauras N, Quigley CA, Rubin K, Sandberg DE, Sas TCJ, Silberbach M, Söderström-Anttila V, Stochholm K, van Alfen-van derVelden JA, Woelfle J, Backeljauw PF; International Turner Syndrome Consensus Group. Clinical practice guidelines for the care of girls and women with Turner syndrome: proceedings from the 2016 Cincinnati International Turner Syndrome Meeting. *Eur J Endocrinol.* 2017 Sep;177(3):G1-G70. doi: 10.1530/EJE-17-0430. PMID: 28705803.

Dicken, BJ, Billmire, DF, Krailo, M, Xia, C, Shaikh, F, Cullen, JW, Olson, TA, Pashankar, F, Malogolowkin, MH, Amatruda, JF, Rescorla, FJ, Egler, RA, Ross, JH, Rodriguez-Galindo, and Frazier, AL. Gonadal dysgenesis is associated with worse outcomes in patients with ovarian nondysgerminomatous tumors: A report of the Children’s Oncology Group AGCT 0132 study. *Pediatr Blood Cancer* 2018 Apr; 65(4): 10.1002/pbc.26913.