

# Hepatic adenoma is associated with Turner syndrome



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### BACKGROUND

- Hepatic adenomas are rare, epithelial tumors of the liver. Typically, they are benign, but are associated with hemorrhage and malignant transformation.
- Turner syndrome is known to be associated with liver abnormalities, but not hepatic adenoma specifically.<sup>1</sup>
- Two index cases of hepatic adenoma in Turner syndrome patients here at UW Health raised the question of whether or not the two diagnoses are associated.

# METHODS

- ICD-10 codes were used to identify patients at UW Health with a diagnosis of Turner syndrome encountered between 2006 and 2020 in the electronic medical record.
- Included patients with:
- Classic Turner syndrome
- Mosaic Turner syndrome (including men)
- Chromosome X deletions with features of Turner syndrome
- Manual chart review of each individual patient was conducted to determine their history of receipt of abdominal imaging (ultrasound, CT, MRI) that evaluated liver parenchyma, and if any lesions consistent with hepatic adenoma were identified.
- Additionally, we collected information on the management of hepatic adenoma once diagnosed (biopsies, surgical intervention), any complications (e.g. rupture of hepatic adenoma), and patient history of estrogen supplementation.

Three of 233 patients with Turner syndrome were found to have hepatic adenoma.

Two were diagnosed incidentally, and one patient had hemorrhagic rupture of hepatic adenoma requiring urgent surgical intervention.

## RESULTS

Turner syndrome:	N = 233 (231 confirmed, 2 unconfirmed potential diagnoses)
Sex:	Female = 228 (97.9%), Male = 5 (2.1%)
History of abdominal imaging?	Yes = 78 (33.5%), No = 155 (66.5%)
Age at abdominal imaging:	Range = 0 – 67 years; Median = 30 years
Hepatic adenoma on imaging?	Yes = 3 (3.8%), No = 75 (96.2%)
Age at diagnosis of hepatic adenoma:	1 month; 24 years; 31 years
Complications of hepatic adenoma:	N = 1 (hemorrhagic rupture requiring urgent surgical intervention)
Patients on estrogen supplementation:	N = 139 (59.7%)

Right: Hepatic ultrasound still frames demonstrating hepatic adenoma measuring 8.81mm x 7.19mm.



### CONCLUSIONS

- The prevalence of hepatic adenoma our population with Turner syndrome was 1.3%, compared to a rough estimated prevalence of 0.04% in the general population<sup>2</sup> (with incidence of about 1:1,000,000 in nonusers of oral contraceptives (OCPs), and 4:1,000,000 in OCP users<sup>3</sup>).
- These findings support that there is an association between Turner syndrome and hepatic adenoma.
- Surveillance hepatic imaging may be beneficial in this population.
- Further evaluation of factors contributing to hepatic adenoma is warranted, including exogenous estrogen exposure.

## ADDITIONAL KEY INFORMATION

#### Citations:

- 1. Roulot D. Liver involvement in Turner syndrome. Liver Int. 2013 Jan;33(1):24-30.
- 2. Kaltenbach TE et al. Prevalence of benign focal liver lesions: ultrasound investigation of 45,319 hospital patients. Abdom Radiol (NY). 2016 Jan;41(1):25-32.
- 3. Patacsil SJ et al. A Review of Benign Hepatic Tumors and Their Imaging Characteristics. Cureus. 2020 Jan 29;12(1):e6813.

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