

Table 1 (abstract 196):

Rate of liver function test elevation after elexacaftor/tezacaftor/ivacaftor treatment according to pretreatment values.

Pre-treatment LFT values (number of patients)	LFT	No. of events	Rate (No. of events per 100 patient-month) [95% CI]
< ULN (n= 38)	ALT elevation (>ULN)	18	9.7 [5.7-15.3]
	GGT elevation (>ULN)	7	3.8 [1.5-7.8]
	Total bilirubin elevation (>ULN)	14	7.5 [4.1-12.6]
> ULN (n= 18)	ALT increment above pre-treatment value	9	9.6 [4.4-18.2]
	GGT increment above pre-treatment value	13	13.8 [7.4-23.6]
	Total bilirubin increment above pre-treatment value	17	18.1 [10.5-29.0]

CI: Confidence Interval. ETI: Elexacaftor, Tezacaftor and Ivacaftor. LFT: Liver Function Test. ULN: Upper Limit of Normality.

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Rare prevalence of colonic sessile serrated lesions in people with cystic fibrosis

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Background: People with cystic fibrosis (PwCF) have higher rates of early colon polyp development and progression to colorectal cancer than the general population, especially those who have undergone solid organ transplantation. In response, the Cystic Fibrosis Foundation developed colorectal cancer (CRC) screening guidelines in 2017 recommending initiation of earlier first screening colonoscopy and shorter re-screening intervals than in the general population. The exact mechanism for how PwCF develop more polyps and whether different polyp types occur at similar prevalence rates as the general population is unclear. Recent studies suggest that sessile serrated lesions (SSLs) have a prevalence in the general population of 8.2% [1], but a detailed review of prior literature suggests that PwCF may have different polyp type distributions [2]. We developed a database of colonoscopy results in PwCF to better characterize the prevalence of SSLs.

Methods: In this retrospective chart review, patients were identified through the University of Washington Cystic Fibrosis Foundation Patient Registry. Inclusion criteria were diagnosis of CF, and exclusion criteria were age younger than 18. Patients' electronic medical records (EMRs) were accessed using the University of Washington Epic EMR system, including linked archival data systems to access partner health system data on the selected patients. The colonoscopy reports were systematically reviewed for selected variables including patient age at time of procedure, date of colonoscopy, bowel preparation quality, cecal intubation, polyp quantity, polyp size, polyp location within the colon, and whether cancer was present. Statistical analysis was performed to compare data with that of the general population.

Results: Seven hundred fifty-seven patients were identified from the CF registry, of whom 170 had available records of prior colonoscopy for any indication; 52.9% were women and 47.1% men. Mean age at time of colonoscopy was 43.2. From these patients, 288 colonoscopy reports were reviewed, and 270 polyps, of all types, were identified. Two patients were found to have solitary SSLs (Table 1). SSL prevalence was 1.2%, significantly lower than what has been described in the general population ($p < 0.05$). There were 41 colonoscopies in our database from before 2010, from which 14 polyps were found; none of these polyps were hyperplastic or other serrated polyp types.

Table 1.

Characteristics of two people with cystic fibrosis found to have a sessile serrated lesion (SSL). CRC = colorectal cancer; CFRD, cystic fibrosis-related diabetes.

Variable	Patient 1	Patient 2
Year SSL discovered	2016	2015
Age at colonoscopy	>50 years old	>50 years old
Polyp Location	Right Colon	Right Colon
Size of SSL	<5mm	<5mm
Lifetime total polyps	13	15
Polyp on first colonoscopy	Yes	No
3 or more polyps (any colonoscopy)	Yes	Yes
Sex	Male	Male
Lung Transplant	Yes	No
FEV1 (% Predicted) at time of colonoscopy	39	105
Family History of CRC	No	No
Personal History of CRC	No	No
History of DIOS	Yes	No
Pancreatic Insufficiency	Yes	Yes
CFRD	Yes	Yes

Conclusions: At our center, PwCF had a significantly lower prevalence of SSLs than what has been described in the general population. Patients who developed SSLs appear to follow age-dependent trends typically seen in the general population. Low SSL prevalence may shed light on the tumor biology of PwCF. Low prevalence of SSLs in PwCF may affect the utility and advantage of more-expensive stool DNA tests over fecal immunochemical tests for CRC screening.

References

- [1] JIjspeert JE, de Wit K, van der Vlugt M, Bastiaansen BA, Fockens P, Dekker E. Prevalence, distribution and risk of sessile serrated adenomas/polyps at a center with a high adenoma detection rate and experienced pathologists. *Endoscopy* 2016;48(8):740–6. <https://doi.org/10.1055/s-0042-105436>.
- [2] Niccum DE, Billings JL, Dunitz JM, Khoruts A. Colonoscopic screening shows increased early incidence and progression of adenomas in cystic fibrosis. *J Cyst Fibros* 2016;15(4):548–53. <https://doi.org/10.1016/j.jcf.2016.01.002>.

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Weight and body composition of school-aged children with cystic fibrosis and extended modulator therapy

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Background: Body mass index (BMI) greater than the 50th percentile has long been the nutritional target for children with cystic fibrosis (CF), but increasing numbers of children have become overweight or obese [1].